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CHAPTER CLXVII

THE PHYSIOLOGY OF THE NERVOUS SYSTEM IN EARLY LIFE

By Percy G. Stiles, Ph.D.

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The Modern View of the Nervous System.—If a single word is sought which shall characterize the work of the nervous system the term *coördination* will probably be fixed upon. From an objective standpoint the brain and the cord with their radiating and converging nerve-paths serve to adapt the activities of various organs to conditions affecting other structures. In particular, the nervous system maintains the essential connection between the *receptors* or sense-organs, on which the energies of the environment are brought to bear, and the tissues of contraction and secretion (effectors) which manifest the reaction of the individual to stimulation. Such a statement as this may seem grossly inadequate since it fails to express the rôle of the nervous system as the instrument of consciousness. The criticism has weight; nevertheless it is possible to proceed much farther than might be anticipated without abandoning the external and analytic attitude.

In recent writings we find the word *integration* frequently employed to indicate the sum of the nervous functions. It is well chosen, for to confer integrity, wholeness, or unity upon the associated tissues is evidently the supreme work of the neural elements. The nervous system is the chief agency by which something which in its absence would be merely an aggregate or a colony is elevated to the rank of an *individual.* We need often to be reminded that there is another type of integrative action, the chemical. A given part of the body may influence another at a distance not only through reflex relationship which is maintained between the two but also through the medium of active substances conveyed by the circulation. Interactions of this kind between different organs are now known to have much greater variety and importance than anyone would have asserted a few years ago. In the early stages of development chemical factors must be held responsible for almost all that occurs in the rudimentary organism. The hereditary forces which are shaping the embryo can hardly be conceived of except as chemical peculiarities which differentiate its regions. Peritz[1] makes the striking statement that there are only two influences which, jointly, determine the organization and life-history of the nervous system, namely, *internal secretions* and *use* (or education).

An idea somewhat similar to this has been developed by Noël Paton.[2] As regards the earlier stages of the formative process he distinguishes two periods. During the first of these the guiding influence is that which he designates as "hereditary inertia." During the second the part played by the internal secretions is recognized. By hereditary inertia we must understand the sum of the chemical influences which arise within the developing cells of the system itself. The internal secretions which later begin to exercise control are the products of tissues other than those of the nervous system and are conveyed to it by the body fluids. Noël Paton, like Peritz, points out that these chemical agents operating within the organism are to be supplemented in due course by the impulses arriving from the receptors. It is the development consequent upon these last which Parker[3] has compared to the transformation of a desert into a fruitful land by irrigation. We shall find that it is most difficult to determine how far the evolution of a function is dependent upon the *use* of a complex and how far it is established in the natural and spontaneous perfecting of the mechanism from within.

Attention may be called to the fact that the earlier developmental influences, which we have called chemical, must be fairly uniform for all the members of a species. The later influences, springing from contacts with the environment, will be much more variable. In those cases in which the organization of the nervous system is carried nearly to its limit by the internal or chemical factors we cannot look for marked individuality. There will be little differentiation through experience. Moreover, those animals whose nervous systems are shaped according to chemical rather than environmental influences may be expected to be competent, almost from birth, to lead independent lives. In contrast with them, those animals whose neural development awaits the modifying action of afferent currents must be comparatively helpless at first. Allen[4] calls attention to the fact that the difference is shown when the guinea pig is compared with the white rat. At the time of birth the former is remarkably advanced in all its capacities. The white rat is much less so. The rat is correspondingly more teachable than the guinea pig, which is to say that the nervous system is plastic under the impacts made upon the receptors. Of course the human infant represents the type in which the largest share is left to post-natal conditions. A long period of dependence upon the parents is necessitated and, as Fiske many years ago pointed out, this prolonged opportunity for communicating to the child the acquisitions of the previous generation may have had much to do with the evolution of man.

To appreciate how subtle and delicate are the chemical influences which direct the formative process we have only to consider how the outgrowing nerve-fibers are guided to their particular destinations in all parts of the body. The wonderful precision of their course to appropriate terminations can only be attributed to chemotaxis but the mind is overwhelmed in the attempt to conceive the number and refinement of the differentiating qualities concerned. The studies of Harrison[5] are immensely suggestive.

Only of late years have we appreciated how involved are the relations between activities which we choose to regard as "nervous" and those which we hold to be chemical. Such structures as the pituitary and the adrenal bodies are found to have embryological kinship with the true nerve-elements but they disseminate hormones instead of nerve-impulses. Elliott[6] has discussed this reciprocal alliance. We have evidence of its existence in the mature organism and we must conclude that it is influential at the earliest periods. The adrenal is a compound organ said to consist of a cortex and a medulla. At the time in embryonic life when the brain is most rapidly developing the adrenal cortex is most prominent. In headless monsters the adrenal cortex is said to be wanting (Bullock and Sequeira[7]). During the first three months after birth, the brain being now well formed, there is a marked atrophy of this part of the gland. So distinctly is this the case that the adrenal actually diminishes in size, despite the fact that its medullary (chromaffin) portion is on the increase (Park[8]).

The Growth of the Nervous System.—A great mass of data can be found relating to this subject. The older findings are incorporated in Donaldson's book.[9] It will be sufficient to emphasize a few salient facts. It is obvious that the brain at birth is disproportionately large as judged by adult standards. From birth to maturity the mass of the body increases about twenty-fold. The weight of the brain increases only 3.7 times. Moreover, it doubles in the first year and at the age of seven it has reached fully four-fifths the final maximum. We are at once brought to appreciate that organization is far more significant than simple growth. From the single standpoint of size the brain of the newborn child is advanced beyond any other organ—unless it may be the eye—but among them all it is clearly the farthest from complete functional development.

A question of the greatest interest is the time-limit of cell division in the central nervous system. According to the older accounts no multiplication of true nerve-cells occurs for a month or two before birth and there is no increase in their number during extra-uterine life. Of late there has been a disposition to qualify this view and to admit that some post-natal division of cells in the gray matter may take place. This has been determined to be the case in certain animals like the rat which are comparatively helpless when born (Allen[10]). The evidence rests upon the observation of mitotic figures in sections of the cerebral and cerebellar cortex. Whether we attach any importance to the division of the nerve-cells after birth or not it is substantially true that the great advances made by the human subject from birth to the prime of life are dependent upon the establishment of functional relations between existing cells rather than upon an increase in the number of these units. Another consideration must be borne in mind: While actual cell division may be too limited to be of material importance it is believed that vast numbers of cells remain for years in a state of arrested development, at last to assume the character of active elements in the cord or brain. It is in this way that we account for the figures obtained by Kaiser (cited by Hall[11]). This investigator computed the number of cells in the cervical enlargement of human spinal cords, comparing specimens from subjects of different ages. The number of fully developed cells apparently doubled between birth and the age of 14. The interpretation placed upon this appearance has been that minute cells, for a long time incapable of functioning, have at length gained the full rank of their fellows.

A glance at any reliable plates made from sections of the cerebral cortex (for example, those of Bolton[12]) will show that even when we make all possible allowance for cell division and the late maturation of inconspicuous elements in the system the increase of typical cells by no means keeps pace with the spatial expansion of the tissue. As the process of growth goes on the intervals between neighboring nerve-cells definitely widen. The texture of the cortex seems to become more and more loosely woven. There is more and more room for the elaboration of the dendritic part of the neurons. Similarly, when the brains of different species are compared it is found that the cortical cells are more widely separated in the higher than in the lower forms.

Myelination.—Many years ago it was pointed out by Flechsig[13] that we may learn much concerning the progress of development in the nervous system by determining at what periods the various fiber-tracts acquire their myelin sheaths. We may assume that it is only as these sheaths are produced that the fibers become functional. A cross-section of the spinal cord at birth shows that all its conducting bundles are apparently matured save for one important exception. This is the "Pyramidal Tract" in the lateral column, together with a few fibers of associated function in the ventral region. There is little doubt that the fibers of the pyramidal tract are those which will later transmit the descending impulses which result in voluntary movement. The late maturing of these paths supports the view that the muscular reactions of the newborn are mediated by other tracts. Rachford[14] refers to the immunity of very young babies from the convulsions which are so common a little later. This shows, as he believes, that the cortical motor system remains at first in abeyance.

In the cerebrum the sequence of myelination has proved especially suggestive. According to the standard description the fibers in this part of the brain may be assigned to two categories. There are those which unite the cortex with lower stations, either conducting impulses downward or conveying them from inferior levels to the cerebral gray matter. These are the projection fibers, efferent and afferent. The pyramidal fibers, just referred to, are conspicuous (efferent) members of this class. Among the afferent projection fibers are those which connect the optic thalamus with the post-central and other portions of the cortex, the optic radiation from the lower visual centers to the cuneus, and many more.

The elements of the second order in the white matter of the cerebrum are the association fibers. Their distinguishing mark is the fact that they begin and end in the cortex. They may be very short, link-ing adjacent convolutions, or they may unite the most remote localities. We may properly place in this class the commissural fibers of the cerebrum—or at least the majority of them—since they conform to our definition; we may, however, prefer to make a distinct class for those fibers which cross from one hemisphere to the other. It was the contention of Flechsig that the projection fibers gain their myelin sheaths earlier than the association fibers. His elaborate diagrams, designed to show how the cortex may be subdivided into areas of primary and secondary maturing have been widely reproduced. Parallel studies by Brodmann[15] have led to conclusions which are somewhat at variance with those of Flechsig. But the chief contention seems to be sustained: that the first cerebral mechanisms to be developed are those which put limited portions of the cortex in functional connection with centers below, the provision for the interaction of the various cortical regions being a later and a more gradual evolution.

Inferences based on the order of myelination have been supplemented by studies of the development of the cortical strata, notably by Bolton[12] and Campbell.[16] It is impossible here to proceed far with a subject of such difficulty. A few general statements may be made. The minute structure of the cortex presents local variations which it is of the utmost interest to interpret. At least five layers can usually be recognized and their probable significance can be inferred from the temporal order of their development and their relative prominence in regions of known function. The five layers referred to are commonly called (1) the molecular or outer fiber lamina, (2) the pyramidal-cell lamina, (3) the granular or middle-cell lamina, (4) the inner fiber lamina, (5) the polymorphic-cell lamina. Comparative anatomy and ontogeny unite to support the view that the inner layers are the primitive ones. In the mature human cortex the inner fiber lamina is probably the chief source of motor discharge while the granular stratum is receptive. The two layers external to the granular are assumed to have associative and, therefore, higher functions.

Evidence in support of these views is found in the following facts: (1) That the large Betz cells, giving rise to the fibers of the pyramidal tract, are found in the inner fiber lamina, (2) that the granular layer is remarkably developed and, one may say, reduplicated in the "visuosensory" area, (3) that the molecular and pyramidal-cell laminæ are most elaborated in the traditional association fields. They are definitely imperfect at birth in the pre-frontal region. The trained observer discovers appearances in these areas up to the eighth month which denote incomplete organization and a non-functional state (Peritz[1]). Bolton has seen in the brains of subjects who have become demented late in life a reduction of the surface layers which strongly suggests a return to the infantile condition.

The conceptions we have just reviewed are well adapted to support a psychological exposition of infant life. We may assume that at birth the impulses from the sense-organs find their way to the primary cortical stations but do not radiate extensively from these regions.

The corollary with regard to consciousness may be supposed to be an experience of sensations which are devoid of suggestiveness. Furthermore, there can hardly be any inferences as to the origin of visual, auditory, and tactual impressions from common sources. Thus Gordon[17] has analyzed in a manner at once brilliant and sympathetic the way in which the mother enters into the thought of the baby. At first she is represented by a recurring patch of color, a soothing sound, and a comforting touch. To realize that these are merely three manifestations of an entity, a "benign power," is to take a momentous step in the mental advance. We may well suppose that the physical concomitant is the establishment in the cortex of association paths, uniting the primitive sense areas through intermediate zones.

The suggestion of Bolton,[12] referred to just above, to the effect that degenerations of certain types occurring in later life may partially reproduce infantile conditions is very interesting. It may sometimes apply to structures below the cortex. For example: We have the observations of Head and Holmes[18] respecting the symptoms which accompany lesions of the optic thalamus. In some of the cases described cutaneous sensations have been curiously intensified and given command of the attention. Experiment shows that this is not due to a lowering of threshold but to an exaggeration of affective tone; pleasantness and unpleasantness alike are raised to the highest degree of vividness. There is something reminiscent of childhood in this. The condition of the thalamus and its connections before the completion of organization in early life may well be one to which reversion by pathologic processes can sometimes take place.

Some writers have asserted that the cerebrum at birth is practically without influence upon the reactions of the child. This seems to be approximately true, as may be gathered by considering the record of a remarkable case, the acerebrate subject of Edinger and Fischer.[19,58] This child died in 1913 at the age of nearly four years. It was found post-mortem that the hemispheres were totally lacking, the space being occupied by sacs of fluid. For about three weeks after birth this baby was regarded as normal, though it was, perhaps, not seen by the most competent observers. Its suspicious stillness, fixed eyes, and slight general rigidity, then attracted attention. The reflexes of sucking, swallowing, and other actions connected with the digestive tract were satisfactory. The eyes were closed to bright light and the child started at loud noises. The hands were not clasped upon objects placed in them; here we may note a definite defect as compared with the normal newborn. The changes observed in this subject with the passing of time were but slight. There was no gain in muscular movement. During the first year there was no loud crying, only a weak whining sound. Later the cry became loud and persistent. It could be stilled by pressure upon the fontanelle but was a serious annoyance to the household throughout the remainder of the child's life.

The Capacities of the Newborn.—Before we enter upon a consideration of this topic it will be well to say something of the classification of nervous reactions. This has been ably treated by Hough.[20] The disposition has been to make the most of the reflex conception in all our nerve-physiology. This leads us to seek an external cause for every manifestation of activity on the part of the skeletal muscles. We must remind ourselves that some movements are certainly in another class. An act should not be regarded as a reflex if the condition which occasions it arises within the central nervous system. Our breathing is the foremost example of a movement of which this is true, a type of action commonly called automatic. The stimulus in this case is provided by the carbon dioxide tension (or H-ion concentration) at the respiratory center. The taking of the first breath may be prompted by stimuli applied to the skin but the fundamental cause of the succeeding movements, if not of the very first one, is central rather than peripheral. What shall be said of the movements of the fetus? Morat[21] has expressed the opinion that they are reflex, conveying the impression that they are due to pressure brought to bear upon the body of the unborn child. Sometimes they may be asphyxial, in which case their origin will be central.

It is a common practice to describe the aimless movements of newborn babies by the term "impulsive" or an equivalent (Mumford;[22] Compayré[23]). The idea underlying the description is that excitation at the centers occurs as an incident of the chemical processes of development. Motions from such a cause must be quite accidental and it is not clear they serve any useful purpose save as they make more easy the transmission of effects from the nerves to the muscles. We have an interesting body of evidence indicating that this is more difficult at first than it will be later. Holmes[24] has determined the strength of the galvanic current necessary to excite contractions of muscles in children, using convenient motor points so that the stimulation is by way of the nerve, and has found that the thresholds are distinctly high. If the value of current required at one month is represented by 5, the value at six months will be about 4, at one year about 3, at two years 2.5, and at four years 2. It would have been freely predicted that the neuromuscular thresholds in infancy would be found low.

The reactions of the newborn have been recorded by many writers. Studies of individual children have furnished material for a number of books to some of which we must presently refer. For a general survey it is convenient to make use of such collective observations as those of Peterson and Rainey[25] and the recent paper by Blanton.[26] From the latter source the following statements may be transcribed. Among the coördinated acts performed by infants *sneezing* has been noted from the first minute, *yawning* at five minutes, *hiccups* at six hours, and—naturally—sucking, swallowing, crying, micturition, and defecation from correspondingly early hours, the last two probably being executed before birth. (This has been judged to be the case from the condition of the amniotic fluid.)

The crying of babies in the first days has been quite carefully investigated by Blanton as well as previously by Borgquist.[27] The quality of the cry is said not to vary much with the stimulus which occasions it except that the pitch is raised extraordinarily when colic is the cause. Sobbing is not heard at first; it begins, according to Borgquist, in the second month. Crying is sometimes accompanied by the shedding of tears even in the first weeks (Blanton) but tears are not usually conspicuous before the third or fourth month. Preyer[28] in his classic work stated that the crying of very young babies was not attended by the woeful expression which later reinforced the effect of the demonstration. This is contradicted by Blanton.

The eye-movements have been subjected to much scrutiny. There are many references to a temporary asymmetry in the direction of the axes. After a few weeks this is noticed less often than a marked convergence which probably bears no relation to the child's germinating interest in its surroundings. The eyes are said to be fixated by bright objects and to follow them slowly in the first week. The protective lid reflexes are rather sluggish at the outset. Miss Shinn,[29] in her remarkable book which must often be quoted, records the closing of the eyes against bright light on the twenty-fifth day and a slow wink when the eye was threatened on the fifty-sixth.

The capacity of the newborn to hang by the hands from a rod has been much discussed. The original observation is attributed to Robinson.[30] Blanton has amplified our knowledge of this unexpected power. According to this author the reaction is most easily demonstrated upon excited infants but it is found to be preserved almost to the last by dying babies. A child of 52 days supported her weight *with one hand* for 42 seconds. The ability is not retained beyond the third month if so long as this.

The respiratory reflexes seem to be among those most completely provided for in the earliest period. It would be of interest to know whether the threshold of the respiratory center is markedly different in babies from what it is to be later. A personal communication from Higgins of Johns Hopkins is to the effect that the carbon dioxide tension in the alveolar air of infants is definitely less than for the adult. The difference is about 10 per cent. But the investigator does not feel sure that the center itself has a greater sensitiveness to this gas or to the H-ion concentration of the blood than it will have in later life. There are incidental factors which leave the matter difficult to determine.

Babies often illustrate the phenomenon known as *apnea*. This is a suspension of the breathing movements following a period of excessive pulmonary ventilation. It is caused by the lowering of the carbon dioxide tension (acapnia) consequent upon the unusual diffusion of this gas from the blood. The normal stimulus to the respiratory center is temporarily lost. It will be restored as soon as the general metabolism has renewed the carbon dioxide in the circulation but in the meantime the oxygen store will have been drawn low and under some conditions the oxygen lack may become serious before breathing is resumed. A baby after violent crying may cease to breathe and become so cyanotic as to excite alarm. The case is apparently just that which has been outlined: The crying constituted an over-ventilation of the lungs and depleted the carbon dioxide of the blood. The temporary stimulus which caused the crying having ceased to operate, the normal stimulus for breathing is wanting and an interval of apnea ensues. The darkening of the blood is the sign that oxygen is being consumed while the breathing mechanism remains at rest, awaiting the rise of the carbon dioxide to its ordinary level.*

By contrast with the well-developed respiratory equipment the coördinated system for the maintenance of a uniform-body temperature is at first decidedly imperfect. The ease with which babies are chilled is familiar. For example: Budin[31] reports the survival of babies whose rectal temperature has been depressed even to 32°C. It is, of course true that for any small organism the surface is strikingly extensive in proportion to the mass and heat-loss tends to be correspondingly free. This is the more so on account of the highly vascular skin. The hands and feet of young babies may become very cold to the touch without exciting any obvious protective reflex and without seeming to cause discomfort. The uncertainty of temperature regulation is shown also in the development of fever when the conditions inducing it seem trivial (Copeland[59]).

The alimentary canal of the child is probably characterized by more or less marked hyperactivity. Hess[32] has been led to this conclusion. He calls attention to the gastric movements often visible through the thin abdominal wall. The observations of Carlson[33] are of particular interest. He has made graphic records of the contractions executed by the fundic pouch of the empty stomach in subjects of all ages. Such contractions are known to be associated, probably in a causal sense, with the pangs of hunger. He has inserted tubes with soft rubber balloons attached into the stomachs of infants—30 in all—and has demonstrated that the hunger contractions are frequent and intense. This is just what might be inferred from the crying of hungry babies. The work of Carlson has been confirmed and extended by Taylor.[34]

Peritz[1] has ventured the generalization that the newborn child is "sympathicotropic" and is destined to develop into a "vagotropic" condition. The most evident fact in support of such a claim is the progressive reduction of the heart-rate. The slowing of the rhythm is supposed to indicate the establishment of vagus inhibition.† Experiments on young animals are said to show that the vagus has, at first, no efficient connection with the organ which it will later bring under restraint. It may be questioned whether this shows that the heart is under a sympathetic (accelerator) influence at first; perhaps it is merely left free from any type of nervous government and is giving expression to its own inherent tendency. Certainly the active alimentary canal of the infant does not suggest a sympathetic domination for this division of the autonomic system has an inhibitory effect upon most of the digestive apparatus. A more natural inference is that none of the pathways from the central nervous axis have yet gained command of what is, at the outset, a series of self-directed organs. Taylor[60] has shown that the secretion of gastric juice in young babies is a rather constant process and probably guided by chemical rather than nervous conditions. The periodic "appetite secretion" has yet to be established. Since the psychic secretion requires the mediation of the vagus fibers this fact supports the view of Peritz that a "vagotropic" condition is to be gradually attained; it does not prove that the initial state is "sympathicotropic."

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* Sometimes such suspension of breathing may be due to spasm of the glottis.

† During the first week the heart-rate *increases* from about 112 to about 126. The decline comes later (Benedict and Talbot[61]).

The cardiac sphincter seems in the first months to have little positive action. Regurgitation from the over-filled stomach is one of the most familiar episodes of infancy. The act is little suggestive of vomiting for there are no convulsive contractions of the diaphragm and abdominal walls. The milk flows quietly through the esophagus, escaping from the tonic pressure of the stomach, and often trickles from one angle of the lips with incredibly little interference with the breathing. It is not likely that any sensation of nausea attends this gentle removal of an excess of food. The incompetency of the guardian sphincter is another sign that central control has not been fully assumed. Continued into childhood this weakness of the cardia may lead to the habit of rumination (McClure,[35] Grulee[62]).

Hymanson[36] has gathered figures regarding the time required for the passage of carmine through the alimentary canal of the newborn. According to his findings this time varies in healthy subjects from 4 to 18 hours. The corresponding interval in the adult is usually assumed to be 24 hours on the average with extremes of something like 10 and 40 hours. The range of variation in Hymanson's figures is so great that it is difficult to comment on them. Four hours seem to indicate a passage that is rapid compared with the average of later life. But 18 hours in a small organism strikes one as long. Of course, the milk diet is rather constipating compared with the mixtures ingested later.

Blanton[26], has recorded a solitary fact concerning the male genitalia in babies. This is the occurrence of more or less complete erection at birth or within a short time. The significance of such a reaction appears doubtful; perhaps it is more correctly described as a deficiency of tone in the vessels than as an active phenomenon.

The Sleep of Infancy.—Sleep, which we are accustomed to think of as a suspension of consciousness, is more profitably viewed in the early weeks from an objective point of view. So regarded, it is essentially a state in which the thresholds are greatly raised, with a consequent reduction of the reflexes and the general tonus of the system. It has been spoken of as a reversion to the condition before birth. Compayré[23] has suggested that it is at first the natural state and interrupted only by strong stimuli such as arise from the hunger contractions. It is, according to this view, *waking* rather than sleep for which we have to account. With increasing irritability of the receptors and provision for more extended radiation of sensory impulses, external contacts serve more and more to keep in progress those reactions which, we say, belong to the waking condition. But effective stimulation leads to fatigue and the thresholds rise—hence, the recurrence of sleep. Compayré accordingly puts it that the early sleep is due to the inadequacy of the means of stimulation while that of the older and more active baby is due to the exhaustion of the receptors. High thresholds figure in both, in the first owing to the immaturity of the structures, in the second to metabolic accumulations. It may be that the newborn child is to some extent drugged by its own metabolites because of restricted elimination during the fetal life.

At first long periods of unbroken sleep are not usual because, within three hours of feeding the hunger contractions are resumed. After the quiet interval has been somewhat lengthened the baby still wakes once in the night and requires to be fed. A rhythm may be impressed upon the nervous system which is only gradually—perhaps never entirely—lost. The child of three, though usually going through the night without waking, yet gives signs of a shoaling of sleep at a certain time. Two periods of profound slumber are separated by a shorter one of restlessness in which a slight disturbance suffices to rouse the sleeper (Czerny, cited by Howell[37]). The disturbances of sleep in infancy and childhood (Strauch[63]) indicate that the depression of the various functions is much less uniform and symmetrical than in the adult. Sometimes there is motor quiet with apparent receptivity of suggestions. The opposite condition of restlessness or somnambulism with marked indifference to external stimuli is also possible.

Early Sensory Capacities.—Parker[3] has reminded us that in the course of evolution reflex responses are early and evident facts of existence while the arrival of sensations is determined with difficulty and is certainly much more tardy. What we are obliged to attend to in the first days of extra-uterine life is the possibility of obtaining reactions to stimulation of the various receptors and inferences with reference to the *feelings* of the baby have to be drawn with caution. This, however, scarcely lessens the interest of the results.

It has been frequently asserted that the thresholds in infancy are characteristically low. This is by no means borne out by most of the data which have been collected. We have already referred to the fact that there is a higher resistance to the effective discharge from the central nervous system to the skeletal muscles than there will be later on. This want of irritability on the motor side has to be reckoned with when we try to form judgments respecting the excitability of the sense-organs yet it seems, on the whole, that these structures and their pathways to the central stations are not abnormally pervious. We shall find it more reasonable to maintain that there is lack of inhibition than that the thresholds are particularly low. The distinction is one which needs to be carefully considered.

It is well to remember the facts demonstrated by Porter[38] regarding thresholds and strychnin poisoning. A casual observer, seeing an animal thrown into tetanus by slight stimulation would infer a great lowering of thresholds. But Porter's painstaking analysis leads to a different interpretation of the condition. He has found that the *minimal* stimulus for the nervous system affected by strychnin has about the same quantitative value as for the normal. In other words, it is not especially easy to induce a reaction. On the other hand, as soon as the threshold is passed the responses are excessive and misdirected. The infant nervous system very probably resembles one upon which strychnin has taken effect: its thresholds are not low but impulses which break through the central resistance rove widely and call many effectors into play. The following analogy may be helpful. A pianist can produce loud tones by striking the keys of his instrument forcibly. An organist gains nothing by extra effort applied to the keyboard; he must chiefly depend upon his stops to add to the volume of the tone. The nervous system poisoned by strychnin is like an organ rather than a piano and it is an organ with all the stops drawn. It is neither easier nor more difficult to press the keys than under other circumstances but every response is of startling intensity.

Under the influence of strychnin impulses which normally make for inhibition come to have excitatory effects. It has already been suggested that the want of inhibition is a leading characteristic of the nervous system in early life. This is signally shown, up to a late stage in childhood, by the rebellion against sleep. Until the need becomes intense all the energies are applied to postpone its onset. Psychologically speaking, the child strives to remain awake, lacks the true sense of its own fatigue, and is as far as possible from courting the sleep which later in life will appear so greatly to be desired. The situation can be expressed objectively by the statement that the central organization which makes for action is far better developed than its counterpoise.

The tradition of low thresholds is upheld by Mercier.[39] It is urged that the receptors must be highly irritable because their stimulation leads to results that seem disproportionately intense. We have offered an alternative to this conclusion but we must not overlook the facts which have seemed to favor it. We have, for example, the provocation of convulsions as the apparent consequence of peripheral causes which would later be reckoned trivial. Among such irritating conditions we may mention, following Holt and Howland,[40] indigestion, parasites, phimosis, adenoids, and, later, eye-strain. There are frequent instances of injuries of a lasting sort from shocks. On the other hand, it is not clear that the immediate reaction to localized stimuli is excessive. The indifference of babies to cold fingers and toes has previously been noted. Blanton[26] has recorded active reflexes in the newborn when alcohol is dropped on the abdomen. It is certainly no less easy to elicit such movements in older subjects. Friction induces lively muscular responses in young babies but Blanton believes that the pain threshold, judged by the effect of a pin-prick, is definitely high.

The sense of taste is said to be demonstrable in the first weeks. Sweetened liquids have been observed to induce sucking movements while bitter fluid has caused grimaces (Kussmaul, cited by Holt and Howland). Peritz[1] tells of a child three weeks old that swallowed buttermilk while refusing sweet. According to general testimony (for example: Shinn[29]) the sense of smell is late in reaching an effective development.

The labyrinth and the associated apparatus in babies must be regarded as difficult to stimulate. It is true that Blanton found the newborn disturbed by a quick downward movement intended to imitate a fall through space. But, as Shinn points out, babies are soothed and quieted by motions which older children would consider rough and disagreeable. One writer mentions the expedient of placing a stick under the rocker of the cradle to give an added jolt to a baby that defied all attempts to quiet it. The result is said to have been all that could be desired. It is noticeable that older children, up to five or six at least, do not seem to find the sensation of dizziness unpleasant. They experiment with it by the most reckless whirling and appear to find amusement in the effect produced. Nevertheless they are often made sick by swinging.

If there is a special sense which really has a low threshold in the early days it is hearing. There is abundant testimony to the effect that the youngest babies are made to start violently by certain sounds. The tones most likely to obtain a response are high-pitched. Shinn noted the reaction on the sixth day when paper was torn. Blanton also found that the rattling of paper was effectual as was the clashing of metal. Perhaps the moist condition of the ear, especially the presence of fluid in the tympanum, makes the organ hypersensitive; it would probably affect an adult in this way. Blanton made a curious trial with the babies under her observation: an attempt to see whether the crying of one child tended to make another cry. This did not seem to be the case, whether the living subject was employed or a phonograph record of a cry.

Among the senses vision has been most studied. We have already referred to some of the ocular reflexes. These are from the very first not only protective but suggestive of the use of the eyes for seeing. Thus Shinn found that on the first day the baby stopped crying when the eyes were uncovered. On the same day they were turned toward a lamp. On the twenty-fifth day the eyes were fixed alternately upon two persons. They followed a moving candle on the thirty-third day; this attainment has often been reported earlier. From the fifth week onward the head was frequently turned with the eyes in such following movements.

Progressive Development.—Thus far we have been considering the inborn capacities of the child and those which are very early acquired. We must now proceed to a synopsis of the gains made in the first decade of life. It is natural to speak first of the motor advances. Among these walking is obviously important. The average ages at which the preliminaries are mastered are given by Holt and Howland[40] as follows: The head is held up strongly in the fourth month. The baby sits securely in the seventh month. In the ninth month it seems eager to stand and does not lack much of the strength necessary to draw itself up upon its feet. At one year it can stand with a slight support, perhaps holding one finger of the attendant. The first steps alone are taken at about 14 months and further progress is very rapid. Of course there are wide departures from these averages.

There has been a good deal of discussion as to whether the movements of locomotion are developed in response to the operation of external stimuli upon the organism or whether there is a gradual spontaneous maturing of a mechanism which in due time provides for their execution. Those who favor the second view have called attention to the fact that movements suggestive of walking (or swimming) may be noticed very early in infancy. Mumford[22] saw these paddling motions on the third day. The legs were generally moved alternately when the child was held free from contacts. Sometimes, however, both arms and legs were moved in synchronous rather than alternating fashion. Graham Brown[41] has described alternating movements of the legs of young animals under anesthesia and has ascribed them to the escape of the motor centers from inhibition, first on one side and then on the other. If we are to believe that walking is attained as a result of the perfecting of a central mechanism taking place independently of external factors, then the child does not "learn to walk" by practice; the awkwardness and non-success of the initial efforts indicate merely that the neural organization has not arrived at that state of efficiency which it will presently reach regardless of its employment. But even if we are inclined to accept this rather unfamiliar view we must reflect that the preliminary exercise is all useful and, in fact, necessary to secure the requisite strength.

In the first year, while steady progress is being made in solving the problems of equilibration and locomotion, a corresponding gain is being made in the use of the hand. The very early clasping of the hands upon objects has been noted. According to Mumford and Buckman[42] the thumb is not used with any effect in these acts. It may be quite left out of the combination at a time when grasping must be regarded as voluntary, the fourth month as set down by Holt and Howland. It is not effectively opposed to the fingers until the sixth month. Shinn reported that the preference for the use of the index finger in poking objects and exploring holes became plain in the tenth month. This is confirmed by Sanford[43] who noted, in addition, that a baby of this age could hold a hair between the finger and thumb, viewing it with apparent interest.

Of course the various gains in motor command are paralleled and indeed conditioned by the development of the afferent apparatus. Only aimless movements can be made without guidance derived from the receptors. According to a convenient classification we may recognize three orders of these sources of nerve-impulses. First, we have the *enteroceptive* nerve-endings, those which lie deep in the visceral tissues and are stimulated by changes incident to local activities. Second, we designate as *proprioceptive* the endings in skeletal muscles and tendons and about the joints, those which are acted upon when tension and pressure in these parts are altered during the execution of movements. Third, we describe as *exteroceptive* all the sensory endings which receive their normal stimulation from without the body.

In the first days of extra-uterine life enteroceptive impulses operate to cause the crying which indicates the occurrence of hunger pangs. Impulses of the same class are sent to the brain as the result of the distention of intestinal loops by gas, the common phenomenon of colic. Still other currents which are to be reckoned as enteroceptive in origin are doubtless passing from the stretched tissues of the lungs up the vagus nerves at the drawing of each breath, the result being a reflex regulation of the breathing movements. The natural tendency is to place in the same category the impulses from the pharynx which lead to swallowing though it may be objected that the surface of the pharynx is, in reality, an infolded portion of the exterior of the body. The nerve-endings in the pharynx are strictly homologous to those in the skin and therefore exteroceptive. Like those in the lining of the nose by means of which sneezing is so readily elicited they are of the same order as the ones in the lips and tongue which have to do with sucking.

The proprioceptive mechanism is presumably brought into play in connection with all muscular movements. Whenever a muscle is contracted impulses ascend from it and from its tendon to the cord and brain. The immediate effect may be a reflex reinforcement of the effort. Under other circumstances antagonistic muscles may be called into action. Sanford[43] states that the early flinging about of arms and legs has its educational value in teaching the child "how movements feel." No doubt this becomes true at some period in development and before the conscious element has risen to importance we may assume that the inflow of impulses from the parts in motion has its share in carrying forward the central organization.

Movements result in the establishment of contacts and so entail the stimulation of the skin receptors. As a baby extends his arm the proprioceptive endings are first subjected to stimulation and when the hand is brought against a surface there is a sudden additional tide of afferent impulses from the receptors for touch and temperature. Sanford has suggested that touch has a peculiarly exciting effect when two parts of the body come into contact, as when the hand reaches the lips or one foot touches the other. In such cases two sets of nerve-currents are started at the same time and if the child has any psychic experience at all the reduplication is novel and interesting. It is superior to the feeling that results from touching an inert object somewhat in the way in which a chord of two notes is more suggestive than either one by itself.

The correlation between visual impressions and manipulation is one of the principal attainments of the first year. The ability to reach and grasp things which have been imaged on the retina is probably not in any degree inborn. It is reported that adults who recover sight after long blindness do not at once recognize the direction, much less the distance of objects. The baby gains this power with surprising rapidity. Shinn observed that her niece rarely bungled in reaching after articles and only in a few cases, after six months, made the mistake of attempting to pick up flat figures. At a certain stage in its career—perhaps at about five months—a child may present a striking picture of intent and concentrated effort as the hand is extended tremulously toward the toy only to pass above or below the coveted treasure. But the unsuccessful trials are comparatively few. "Vision," says Sanford, "is anticipatory touch."[44]

The "mirror writing" which is such a curious perversion in the employment of the hand under visual guidance may be referred to here though it belongs to later childhood. When attempting to make a copy of a word the pupil moves the pen to the left in executing each line that should incline to the right. The result is like a reflected image or the impression on a blotter. The singular fact is that such children, so far as reported, do not make mistakes in reaching for objects. The defect must be assumed to be in a special rather than a fundamental mechanism. It is to be borne in mind that in copying one does not have the eye on the pattern but on the replica at the moment when the lines are being traced. There is a definite incidence of memory in the process though the span is brief.

The unsteadiness of early movements is an obvious characteristic and it long continues to mark the performances of the child, particularly when the small muscles are concerned. Perhaps the final attainment of precision in control of the hand, as demonstrated in drawing figures, may be conditioned by a feature of cortical organization of which Lugaro[45] has made much. This is the "closed circuit" whereby it is supposed that an impulse from a particular cell may set in action a circular current which continually returns, by way of neighboring neurons, to renew the stimulation of the first in the chain. If, at the same time, a discharge along a projection path is proceeding we have here an arrangement singularly adapted to reinforce and prolong the peripheral effect. It is suggested that the development of such circuits is accomplished only by slow degrees and that, pending their establishment, the delivery of impulses from the cortex is fitful and productive of uncertain motions (see also Morat, *loc. cit.*[21] pp. 239, 651).

The closed circuit as a factor in securing steady and effective innervation may not be limited to the interaction of cells which are anatomically close together. It is probable that in the mature nervous system currents having a wide sweep and passing between stations at different levels contribute to the same end. An example is suggested by the studies of Wilson.[46] He has noted that individuals in whom the lenticular nucleus has degenerated become tremulous and uncertain in their voluntary movements. Now the lenticular nucleus is not upon the main path from the cortex to the motor cells of the cord. Any assistance it may render to voluntary innervation must be indirect. Wilson has shown by an elaborate diagram its possible auxiliary service. It is not necessary to enter into details but it is easy to see that if such mechanisms are required for the regulation of muscular movements then any incompleteness of organization in such collateral structures may account, in part, for the unsteadiness of the child.

With the progress of development there is observed to be an increasing command of more peripheral segments. The innervation of the trunk muscles is achieved before those of the limbs are well controlled. Mastery of the muscles of the arms precedes successful management of the fingers. The principle is "from fundamental to accessory." As a matter of fact such an order is inevitable. No set of the smaller muscles can be used with effect until the more massive ones in the background have provided a reliable support. Hall,[11] p. 166, has well urged that this normal progression from basal to accessory muscle complexes needs always to be kept in mind. Harm is done in early childhood by forcing the employment of the latter; by and by the situation is reversed and the danger is that the fundamental muscles will be neglected.

The long continued gain in the command of peripheral muscles is satisfactorily shown in that the maximum rate of tapping with a pencil held in the fingers increases through the whole period from 6 to 16 years. Naturally, too, the power to maintain a high rate improves. Meanwhile the swaying of the body as a whole and the wavering of the extremities when unsupported are diminished. The fact that the basal muscles are in the lead is indicated by the nonchalant skill with which children of 10 or 12 manage their bicycles. At the same age such digital performances as sewing and writing are surprisingly slow and labored.

Vision in Infancy and Childhood.—We are led naturally from a consideration of the eye as a guide to the hand to a more general discussion of the early visual capacities. Something has been said already of the movements of the eyes in the first months. There is no other motor mechanism in which the proprioceptive service is so important as in this one and we may suppose that when the eyes are rolled restlessly in their sockets, with no evident relation to objects of interest outside, the impulses which run centralwards from the active muscles are really contributing to the organization of the brain. It is in no other case so necessary "to learn how movements feel." Judgments of direction and, in a large measure, those of distance depend on the use of the extrinsic eye-muscles and the reception of impulses from them.

Vision, as the adult conceives it, presupposes at least three conditions. The refraction must be good, the retinal organization must be normal, and the central stations must be equipped for their work. To name the third requisite is almost the same as to say that there must be intelligence. In view of these facts it is most difficult to say whether a baby's failure to respond to some ocular test is due to central or peripheral deficiencies. It is often surmised that the child's eyes are at first in focus for near objects but it must be remembered that such objects make disproportionately large retinal images. Perhaps it is the size of the picture rather than its sharpness which commands attention when there are no preformed associations.

The marked convergence of the visual axes which has been referred to may not in all cases signify that near objects are being scrutinized. But before long it comes to have this significance. Peritz[1] states that in the fourth month the convergence begins to be conjoined with contraction of the pupil, the sign of the act of accommodation. From this time on the child enjoys an advantage over the adult in the range of clear vision. Even at 10 years the near-point is only 7.1 cm. in front of the cornea (v. Rohr[47]). Little children often give the impression of being near-sighted when they are merely making use of their easy and extensive accommodation. True myopia is said to be rare in the first years (Hansell[48]).

Numerous attempts have been made to discover whether very young children exhibit discrimination of colors. Parsons[49] has a chapter on this subject which at least makes evident the difficulty of arriving at reliable conclusions. One of the most careful studies is that of Valentine.[50] He recorded the length of time the baby's gaze was fixed upon surfaces of various colors. In such an investigation it is hard to make due allowance for the stimulating effect of simple luminosity and the influence of novelty. But Valentine has felt justified in stating that the color sense is present at three months. Yellow excels all other colors in its power to engage the attention at that age and the order of preference after yellow is white, pink, red, brown, black, blue, green, violet. At seven months yellow is still in the lead while the rest of the list reads somewhat differently.

From a very early period the eye dominates the reactions of the nervous system, at least in the absence of noxious stimuli. Shinn has spoken of the remarkable effect produced upon a baby by the scenery outdoors. This may be exhibited either as excitement or as entranced stillness. Doubtless the luminosity of the environment has much to do with it. Another factor, which some adults may be able to verify by a searching of early recollections, is the amazing display of *parallax*, the shifting of near upon distant planes. This character in the landscape makes a profound impression on the mind of a child and even before he can think about it we may suppose that the ever-varying stimulation of the retina can give rise to nerve-currents of wide influence. When it has become a commonplace in our experience we may still have it brought home to us with something of its original vividness as we watch moving pictures.

We cannot learn much about the vision of the child until he has advanced far enough to give clear indications of recognition or inferential power. Members of the household are certainly differentiated at a surprisingly early period. Shinn was satisfied that her niece accomplished this in the third month. In the fourth month it was noticed that the child was impressed by changes of head-gear. But mistakes as to the identity of associates when seen at 20 yd. were frequent up to the eighteenth month. Whether this means that the eyes are generally accommodated for near vision or whether the ability to interpret small retinal images is lacking is difficult to decide. In the twentieth month this little girl said that a man 25 ft. away was eating when he was really counting money. In sharp contrast to this deficiency the appreciation of pictures was definitely present from the ninth month—another example of the superior effect of near planes and larger images.

As to the auditory apparatus in infancy the statement has already been made that it is readily stimulated. Coördination between the ear and the eye is secured very quickly. The child observed by Shinn turned the eyes and head toward sources of sound with some certainty even at three months and earlier still looked in the direction of the piano when chords were struck. In the sixth month she responded to her name. This is a significant sign of the cerebral evolution. Coming, as it does, long before the first words are framed it is a reminder of the necessity that the auditory centers be well organized before there can be spoken language. Parents must generally have recognized that children have associations with many words—"understand them," as we say—at a time when they have still "no language but a cry." Lovers of animals hold with much reason that the same is true of their pets. In the eighth month Shinn was convinced that her niece knew the members of the family by name. Speech must have its beginning in vocalization stimulated by hearing. A vocal response to piano music was recorded in the case of the same child from the end of the second month onward.

The cutaneous senses in childhood are usually held to be characterized by low thresholds. The thinness of the skin clearly favors the access of stimuli to the receptors and perhaps this explains all cases in which hypersensitiveness seems definitely established. Most of the data have been obtained in pre-adolescent years rather than earlier. The "threshold distance" (for the recognition of two points as separate) is reported to be somewhat, but not strikingly, less at 12 than in the adult.[51] Whether it is really less in proportion to stature seems to be doubtful. Carman[52] found that the pain threshold was a rising one from 10 to 18 years of age both in boys and girls. Presumably it had been still lower in early childhood than at the age when the test was first applied. Expressed in grams of pressure required to call forth a verdict of pain Carman's figures are as follows: For the boy of 10—2253; girl of 10—2191; boy of 18—3267; girl of 18—3077.

The general sensations, those arising from visceral conditions, seem to decline in vividness as the special sensations become dominant. Hunger, which is of such marked intensity at first, becomes much less strong and many children of five or six will not readily leave their play to come to the table. Intestinal pain seems to be much less often felt by healthy children who have passed the first year than it was in the earlier months. As to nausea, the most staggering and appalling sensory experience of adult life, little children usually give way to it only for a very short time before vomiting. Frequently the parent has little reason to anticipate the crisis which is at hand. A partial explanation of the fact that nausea is so little regarded by small children may be found in the excellence of the circulatory system. Splanchnic engorgement in the child is less likely to produce cerebral anemia and faintness than in the adult. We have also to consider the shorter vertical measurements of the smaller body and the resulting limitation of hydrostatic effects upon the distribution of blood.

Are any of the senses brought early to a higher degree of refinement than they can maintain through the next few years? It is in many cases difficult to decide this question because the employment of the sense-organs is at all times conditioned by the central organization with which they stand related. If the brain is advancing in its development while the receptors are retrograding the net result of the partnership may for some time continue to appreciate. Thus Foster long ago suggested that the eye, looked at as an isolated organ, is at its best in early youth when the limitations of intelligence restrict its use to that of a mere toy. Its great contributions to the intellect are made when it is no longer so good an instrument. It is quite likely that some of the simpler sensory judgments are better in pre-adolescence than they are ever to be again. This is asserted by Gilbert (cited by Hall[11]) for the estimation and comparison of weights. This particular ability is reported to reach its acme in girls at 13 and in boys a year later.

The reaction-time of children is long. We may say that this indicates a want of the facility secured by practice; canalization is incomplete. Another way of expressing much the same idea is to say that the nerve-currents are dispersed unprofitably instead of being economically directed toward a definite end. It will be found upon consideration that this is a restatement of the general truth that the young nervous system is distinguished by its lack of inhibition. It is reported of adults, quite contrary to what would be popularly supposed, that individuals regarded as stolid and phlegmatic have the shortest reaction-times. They are highly efficient, so far as this test indicates, because the energy applied is so strictly confined to the precise channels through which the desired result can be obtained.

In this treatment of our subject the point of view has been so far as possible external and objective. By adhering to this method, that of physiology rather than old-fashioned psychology, we avoid much discussion of a dubious and speculative sort. Yet we shall do well to admit some matters which bring to the front the mind and not simply the nervous system of the child. William James has somewhere said that the mental attitude of the baby is expressed by the interjection "Lo!" It is a mood of wonder and surprise at novel sensory experience. We have already suggested the early necessity for associating sensations that have a common origin, a process of education which has its physical equivalent in the opening of tangential paths in the cortex. The bonding of certain sensory with certain motor centers is, perhaps, a process taking place at an equally early period and it soon begins to add to the inborn reflexes new examples of response in connection with which there are impressions of satisfaction or discomfort, willingness or reluctance. Along this line proceeds the transition from action which is obviously reflex to that which we venture to call voluntary—that which is held to exhibit the exercise of choice. Just what are the actual neurological facts described in this facile way is not easy to explain.

A vivid and sympathetic description of the baby's "character" has been given by Sanford.[43] The attention of the child is easily secured but not easily retained. This is to say that it is of the passive type, governed by the strength and novelty of the stimulation and not by any decision to hold a constant mental attitude with reference to the sequence of external events. To say this is to emphasize once more the want of cerebral inhibition. In the nervous system of the child we have the most singular co-existence of plasticity and rigidity. The former quality is shown by rapid habit formation, the latter by the extraordinary resistance to change which is recognized so soon as any action becomes habitual.* Figuratively speaking, we have to do with a brain composed of soft material; it is not only easy to channel its substance but each groove quickly wears to a considerable depth. Moreover, as these metaphorical "ruts" are few in number they are followed with greater regularity. There are not so many intersections or "switches" permitting one type of reaction to pass into another.

This resistance to whatever is new presents itself strikingly in children who are old enough to enjoy stories. Repetition gives them greater pleasure than fresh invention. The introduction of a new episode—sometimes of a new word—in a familiar tale develops a comic and strenuous opposition. This is a symptom of a condition which is profoundly significant. White[53] has, in a most interesting way, developed the thesis that this inertia or conservatism in child nature is a chief source of difficulty in training. It makes every advance laborious. A regressive stress is always present, a desire to revert to what is accustomed and therefore easy. Van Teslaar,[54] in a similar exposition bearing upon the Freudian doctrine, points out that a central problem of early development is to replace the internal sphere of the feelings by the sphere of external relationship. Here, again, we find the child absorbed and satisfied by what has been experienced and requiring the strongest compulsions to make him enter upon new phases of mental life. It is a primitive instance of egotism resisting altruism. If the individual summarizes in his own life the evolutionary history of the race we may see in this refractory attitude of the child the sign of the self-content and non-progressive ancestry that dwelt here thousands of years before him.

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* See Strauch on stereotyped postures in sleep.[63]

This is not the place to attempt an abstract of the Freudian teachings. They belong to special pathology rather than to a general survey like the present. But one point certainly needs the utmost emphasis. We owe to Freud and his school the demonstration that impressions made very early upon the nervous system may confer peculiarities which it will never lose. Their influence, often a disastrous one, may be recognized in dreams, fears, obsessions, and irregularities of conduct through a lifetime. The difficulties of nurture and the responsibilities of parents are found to be even graver than we have appreciated (Holt,[55] Campbell[56]). It is hard to realize the possibility of such enduring impressions when we have constantly before us so many signs that suggest instability rather than retentive capacity. As Sanford says: The baby is "light-hearted but easily moved to tears, affectionate but not loyal, easily angered but not resentful."

At about the age of seven years the massive development of the brain is substantially complete. Many have held that this period is a rather significant epoch. Coming midway between birth and puberty it is suggestive of that stage of more momentous transition which lies so far in the future. It has been noted among the blind that those who have lost their sight before the age of seven have little visual memory. There is a marked contrast between them and others who have become blind a year or two later. Fraser[57] tested a group of blind children in the following way. After each had handled a cube he was asked how many surfaces of the solid could be seen by an observer at one time. The correct answer—three—was generally given by those who had been able to see when eight years old; others showed entirely erroneous conceptions by saying four, five, or six.

Later childhood is a time of considerable competence within the limited sphere of daily experience. The suggestion has been made that it represents a certain plane which the race in its evolution long occupied. There is often a sympathy between the child of 10 and the adults of the family which cannot be maintained through adolescence. The views of such a child are singularly temperate and practical and, in many respects, the scale of values can be characterized as mature. To illustrate this from a piece of excellent fiction reference may be made to Booth Tarkington's "Seventeen." Jane, in the story, maintains easy confidential relations with her mother. Willie's adolescent vagaries are scarcely more comprehensible to his mother than they are to his observant and wondering sister.

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CHAPTER CLXVIII

INFANTILE CEREBRAL PALSIES

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Introduction. —The attempt to group under one heading all the cases suffering as a consequence of destructive lesions of the fetal or infantile brain, is certain to stretch to its utmost the limiting possibilities of that heading. The term Little's disease fails utterly to convey any descriptive meaning to the reader. Infantile cerebral palsy is the best descriptive term we have to apply to this varied assortment of clinical pictures. Destructions occur in the fetal and infantile brain. They are due to a number of different causes. As a result of brain destruction symptoms occur, the nature and combination of which depend upon the location, extent and, also, age of incidence of the destructive process. Since obviously these three factors may vary in innumerable ways, obviously also the symptoms may occur in innumerable combinations. These combinations of symptoms constitute the clinical pictures or syndromes grouped under the one heading of "Infantile Cerebral Palsies." It might be said that infantile cerebral destructions or infantile cerebral anomalies would more correctly include all the conditions since there are cases in which no paralysis, in the ordinary sense of the word, can be demonstrated. In other words the only factor common to all cases is that there is a deficit which has occurred in the developing, pre-adult brain.

Development of Knowledge of Disease.—In the development of our knowledge of cerebral infantile palsy the effort of classification is visible as it is in the development of the knowledge of any other diseased condition. As information accumulated concerning paralysis due to destructions of the fetal or infantile brain, various writers attempted to delineate pathognomonic symptoms, nice differential diagnostic pictures with, in some cases, special emphasis on some particular of etiology, in others, some particular of pathology.

Now that sufficient information has been accumulated it is apparent, or at least it seems so, that any of the common symptoms may or may not appear in any of the different forms, and that any influence that can cause cerebral destruction may have to be considered in the etiology.

Prenatal trauma or infection, syphilitic or otherwise, birth trauma due to difficult or prolonged labor, or to forceps, post-natal trauma, hemorrhage or infection; any one of these may be the cause of the destruction whose clinical manifestations are to be differentiated from each other only with greatest of difficulty.

The first really valuable publication dealing with cerebral infantile palsy was made by Cazauvielh[1] from Salpetriére in 1827. He described under the title, "Recherches sur l'agénésie cérébrale et la paralysie congénitale," practically all of the more prominent features of infantile cerebral hemiplegia. He noted that the arm was more involved than the leg, that contractures and increased tonus were to be found, that the paralyzed extremities were smaller than their opposites, that involuntary movements might be found, that the intelligence was below the average even to the point of idiocy. He mentioned epilepsy though he did not consider it as a characteristic of the disease.

The pathologic foundation for the symptomatology of cerebral infantile palsy was a matter of much argument until Cotard[2] in 1868 announced the results of his investigations. He found four orders of processes. (1) Yellow plaques. (2) Cysts. (3) Atrophy due to absorption of diseased nerve tissue. (4) Diffuse sclerosis. The first three he considered as primary evidences of the disease; the last as at times secondary, at times primary. He considered as the underlying agents to cause these changes, softenings, hemorrhages, trauma and perhaps diffuse lobar sclerosis.

Jendrassik and Marie[3] studied this diffuse lobar sclerosis and showed that it began about the smaller blood-vessels. They also observed that the beginning of the clinical picture of cerebral infantile palsy often appeared during the convalescence from some one of the infectious diseases of childhood, and in these cases they found diffuse lobar sclerosis. Hence, they concluded that it was the result of an inflammatory process originating in the vascular system.

Little[4] had already published his observations on the part that accidents at birth played in causation of infantile cerebral palsy. His work was so thorough, his descriptions of the clinical pictures so clear and exact, his deductions as to the relation of the symptoms to the causative agents so convincing that in English literature infantile cerebral paralysis has been called Little's disease since his time.

In 1885 MacNutt[5] showed that meningeal hemorrhage was the immediate cause of the symptoms in these birth palsies, that prolonged difficult birth entered into the etiology only as it occasioned the hemorrhage.

Contributions from Benedikt[6] and Weir Mitchell[7] emphasized the position of chorea in the symptomatology of infantile cerebral paralysis. Raymond[8] and Charcot attempted to localize the lesion responsible for chorea in the internal capsule but Kahler and Pick[9] recognized that such a narrow view of the pathologic physiology of involuntary movements was untenable.

Epilepsy was mentioned as a symptom by Cazauviehl but its position was established only when H. Jackson developed his views on epilepsy due to a focal lesion of the brain.

Strümpell,[10] in 1885, published his work on Poliencephalitis acuta. He held that sclerotic processes in the brain were, in many cases, due to a specific system disease of the same order as Poliomyelitis. P. Marie[11] recognized the occurrence of inflammatory processes in the brain but, more correctly, maintained his opinion, previously expressed with Jendrassik, that it was not a systemic disease but a vascular disease the exact location of which was more or less accidental.

The improved methods for recognition of syphilis have in later years led to an understanding of the origin of many previously obscure cases. It has been held that the luetic heredity can act in two ways. In one, the more common, the disease process is due to the direct action of the spirochete. In the other, for which we have only negative evidence, the absence of typical microscopic findings, the indirect action of the spirochete is postulated. Due to the action of the spirochete, the mother's blood is impoverished; she cannot give sufficient nourishment to the growing fetus, or going even further back, the egg lacked sufficient germinating power because of the syphilitic disease of the mother.

Also, in later years, the importance of foci of infection in the production of disease in distant parts of the body has thrown a new light on the problem of intra-uterine and extra-uterine infantile brain disease. The conditions which favor metastases in the adult can be at work in the pregnant woman or in the nursing infant.

The foregoing account is taken largely from the article of S. Freud[12] in Nothnagel's System. Since that was written in 1897 much has been written directly about infantile cerebral palsies and much also that bears indirectly on the problems of infantile cerebral palsies.

Miura[13] gathered the evidence seeming to prove the primary nature of microgyri in certain cases. Oppenheim had held that they occurred only as a result of some other process, usually inflammatory. He cites the case of Mya et Levi in which there was no macroscopic change in the brain but which showed a failure of Nissl bodies in the pyramidal cells, their place being taken by a granular detritus. The protoplasmic processes of the pyramidal cells were abnormally short and fine and the axis cylinders were fine and presented poor myelin sheaths. In his own two cases there is no sign of inflammation and he considers the microgyri as primary and the symptoms as due to dearth of Py elements. He quotes Heubner to the effect that two varieties of influences can act upon the undeveloped brain to produce symptoms. One is an inhibiting process of unknown origin, the other some process affecting the brain locally such as hemorrhage, tumor, etc. The inhibiting action may be easily considered as acting most perniciously on the pyramidal cells of the motor cortex since they are called upon to support an unusually long axone and must be able to develop unusual impulsive force to send the necessary stimuli over their long course to the anterior horns of the cord.

In the article on Progressive Lenticular Degeneration in Brain of 1911 S. A. K. Wilson goes into the pathogenesis of involuntary movements and spasticity with characteristic thoroughness. Since involuntary movements are present in such a large percentage of cases of infantile cerebral palsy his work leads a step towards a better understanding of the problems of this condition.

In pure cases of progressive lenticular degeneration there is no involvement of the pyramidal fibers, hence the involuntary movements of Wilson's disease cannot be due to mere interference with transmission of impulses from the cortex down the pyramidal tract. There are mechanisms, however, whose proper functioning is necessary for the elaboration of the impulse before it is sent over the pyramidal fibers. Hence, a lesion of these mechanisms may effect the pyramidal function just as surely as it will be effected by a capsular destruction. One of these mechanisms is the cerebello-rubro-thalamo-cortical tract. The end station of this tract is in the sensory and motor central cortex. Wilson presents evidence which leads him to say that there is no doubt but that in lesions of this corticopetal tract certain involuntary movements are prone to appear.

The work of physiologists and clinicians has led to the conclusion that integrity of the sensory arm of the spinal reflex arc is necessary to a maintenance of muscular tone. Forester's work in cutting the posterior nerve roots, verified this assumption and eventually led him to advise rhizectomy or posterior root section as a means for overcoming the rigidity of infantile cerebral palsies which in some cases seems to be the chief obstacle to adequate functioning.

There has been but little written of the part played by the undeveloped state of the cerebral nervous system at the time of onset of the disease in the production of symptoms which differ from symptoms seen in the adult cerebral palsies.

There are at least three ways in which these differences may arise. The infantile skull is distensible and so will allow of spread of hemorrhages or swelling of inflamed tissue, which would cause death by compression in the adult. Hence, symptoms may arise in infants in cases where death would prevent their development in the adult. The lesions may bring about conditions which in themselves prevent the formation of the multitudinous connections between the different cellular structures of the central nervous system. Cells may be prevented from sending out their usual number of axones and dendrites by the presence of inimical influences. And lastly, whether based on the second premise or not, habitual connections have not been established in the infant as they have in the individual of 10 or more.

The fetus and the newborn infant make many movements before cortical control is established through complete development of the pyramidal tract. As time goes on cortical control over certain parts of the body becomes more necessary to its adequate functioning whereas other parts seem to need cortical control much less. For instance the arm and the lower face perform in obedience to cortical commands a much greater proportion of the time than do the upper face, the trunk muscles and the leg, and so in the adult when a paralyzing lesion affects the cortico spinal tract it is essentially these cortical appendages, the lower face and the arm, that are affected and it is the upper face, the trunk muscles and the leg that tend to regain some of their function.

It has been held that the upper and lower face are apt to suffer equally in the infantile forms of paralysis; also it is well known that paralysis of the legs or at least interference with their voluntary control may be much more marked in the infant in proportion to the arm involvement than it ever is in the adult. This may be due to lesions destroying the paracentral cortex on the two sides. And it may be true that lesions in this particular region are especially liable to occur in the birth injuries due to tearing of the veins at the point of entrance into the longitudinal sinus. As I say, these facts may account for the difference in numbers of incidents in the two ages, but it is almost unheard of to get a simple paraplegia in the adult brain injury. I think it may be that just as cortical control is never nearly so fine in the legs, feet, and toes, as it is in the arms, hands, and fingers, so when interference with the development of this control occurs before any establishment of habit has taken place the legs may suffer more than the arms. For example in a case of diplegia the only voluntary control was over speech and movement of the index finger on either side. It seems like a decided stretch of the imagination to postulate the bilateral escape of cortical centers for the index finger and pharyngeal, tongue and lip muscles. In fact in this cases there was meningeal thickening and edema with atrophic underlying gyri of the entire frontal and central portions of the brain. This diffuse superficial destruction allowed of a certain amount of cortical motor control. What more natural than that it should be directed towards those organs most frequently employed in expressing cortical impulses?

Classification.—The cases of infantile cerebral palsy, showing as they do a great variety of forms, require a classification. It is impossible to consider separately each individual collection of symptoms; they must be grouped under definite headings and the variations from the ordinary forms given their proper valuation.

The causative lesions of infantile cerebral palsy may produce easily classifiable clinical types. This is especially true when there is well marked loss of the power of voluntary movement. On the other hand, a symptom such as epilepsy, idiocy, or involuntary movement, because its nature is such as to place it in the front of the picture; may give an entirely different appearance to the clinical syndrome. This is especially true of those cases in which the loss of voluntary movement is not of high degree.

The name "Infantile Cerebral Palsy" demands a paralysis of the spastic or upper neurone type. In most cases there is both paralysis and spasticity, but the paralysis may be greatly overshadowed by a combination of two factors: The paralysis may be minimal, and the concomitant symptoms may be so striking as to eclipse completely the motor weakness. There are cases in which the paralysis, if it ever did exist, has entirely disappeared, leaving behind chorea, epilepsy, or mental enfeeblement.

In a clinical classification the paralysis comes first. The cases in which the voluntary paralysis is the predominant symptom are the hemiplegias, the paraplegias and the diplegias.

Such a classification refers only to the part of the body whose power of voluntary movement has been affected. It takes no cognizance of the etiology or pathologic physiology. And because of the title, "Cerebral Palsy," it is understood that in all cases the paralysis is due to lesion of the brain. There are exceptions, to be spoken of later.

There are many and large areas of the adult brain that can be destroyed without inherently affecting voluntary movement. The same fact is true, perhaps in even a greater degree, of the infantile brain. Hence the hemiplegias, the paraplegias, and the diplegias, i.e., the various forms of disturbance of voluntary movement do not include all the cases of infantile cerebral paralysis.

To the plegias, therefore, must be added the cases of destructions of the brain which show their presence by intellectual defects, epilepsy, or chorea.

The intellectual grade of these defectives may vary all the way from the lowest grade of idiocy to the high grade imbeciles. Indeed the degree of mental enfeeblement or backwardness may be so slight as to be unsuspected as to its origin. The child of violent temper who later grows up as an incorrigible, a recidivist or a moral pervert may owe his anti-social mentality to some small brain destruction in infancy.

The position of the symptom epilepsy in the symptomatology of infantile cerebral paralysis is as yet largely conjectural. There is no doubt but that epilepsy is much more frequent in infants affected by infantile cerebral paralysis than it is in those who are not so afflicted. It can even be said with considerable assurance that it is most common in the hemiplegias, i.e., in the cases that have developed during extra-uterine life. All cases of post-natal infantile cerebral paralysis do not develop epilepsy by any means. However, that epilepsy is a well-recognized symptom of infantile cerebral paralysis is without question. Indeed it is highly probably that some cases of so-called idiopathic epilepsy are in reality infantile cerebral paralysis, the epilepsy being the only remaining symptom.

Involuntary movements, as a symptom of infantile cerebral paralysis are so striking that they easily put into a class by themselves, the cases in which they occur. So added to the other forms of infantile cerebral paralysis we have the choreic forms. The involuntary movements may be but slight in comparison to the paralysis, the deformities, or the mental enfeeblement, or they may be the only apparent evidence of the destruction of brain tissue. Where they occur, however, some mental retardation and, at least, some clumsiness of voluntary movement, will also be found.

This clinical classification is determined by the predominant symptom in each of the different types of cases. The number of these prominent classifying symptoms present in each case varies greatly. Many cases show them all; others show only one, especially is the latter true of the epileptic, though in practically all of these cases some variation from the normal will be revealed in a careful physical and mental examination.

The cases of infantile cerebral paralysis may be classified also according to (1) the period of onset; (2) the underlying pathology.

I. The period of onset in infantile cerebral paralysis undoubtedly has a marked influence in determining just what effect a pathologic process is going to have upon the brain. Perhaps this is best illustrated in the case of meningeal hemorrhage, which typifies all those cases in which there is a sudden increase in the contents of the skull cavity. The physical conditions inside the fixed walls of the older infant's skull are decidedly different from those within the membranous walls of the fetal or within the semi-rigid walls of the new born. In the younger skulls adjustments can be made that will prevent death from occurring as it surely would occur if such adjustments were not made. Also various transcortical tracts, association paths which are not myelinized until about the third year of life may be greatly hindered in their development in case the pathologic process antidates their establishment. The cases of infantile cerebral paralysis may therefore be divided into the:

(*a*) Prenatal or intra-uterine.
(*b*) Birth palsies.
(*c*) Acquired or post-natal.

II. It is important to classify the infantile cerebral palsies according to pathology. This is difficult for several reasons. The cases of prenatal onset or origin are to be seen only after birth, hence the acute manifestations may have passed into the chronic and reparative, when the examination takes place. Changes in the brains of individuals living a long time after the onset may be very difficult of interpretation.

All cases may be divided into four classes so far as the pathologic basis is concerned. It is possible that one of these, namely, hereditary, or congenital, maldevelopment, is more correctly included under one of the other three—one of them may be the cause of the maldevelopment.

1. Inflammation.
2. Hemorrhage or thrombus.
3. Trauma.
4. Congenital maldevelopment.*

These three or four general pathological processes underlie the more concrete forms that are found at the post-mortem table at a period more or less distantly removed from the onset.

A classification such as that given by Sachs[14] in 1895 is still useful.

Inflammation includes both the acute and the chronic inflammatory processes. Among the acute inflammatory agents are those which affect the brain as a complication of the acute exanthemata, gastro-intestinal disturbances or acute inflammations of the respiratory tract. Syphilis and tuberculosis are the two chronic inflammatory processes which more commonly find lodgement in the brain.

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* Embolism is included under the second heading; the embolus produces its action as does the thrombus by closing the lumen of the vessel.

In many cases there is an element of both number two and number one. Emboli containing pathologic germs may plug up cerebral vessels and set up inflammatory processes. If the vessels are very small and the emboli numerous, a diffuse sclerosis will develop. If the embolus is larger and solitary then an abscess is the more probable outcome, unless the embolus be of tuberculous or luetic origin, in which case either a tubercle or gumma develops.

The fundamental process may also partake of both number two and number three. Trauma, as a matter of fact, usually does much less harm directly than indirectly through the hemorrhage occasioned thereby. The hemorrhage may exert its action quite locally or may spread out over a rather large cortical area and of course it may be quite large or quite small. In the former case it may even go so far as to compress greatly the hemisphere over which it develops.

The congenital maldevelopments are of all degrees of intensity and extension. It is a simple matter to say that they arise as the result of some defect in the keim, but it is as yet unknown whence arise these original defects. They may be due to some inherent weakness of development, some lack of force to bring about multiplication of cells at the right time, or there may be inflammatory or hemorrhagic processes in the young fetus, all the original traces of which are removed or out-grown.

CLASSIFICATION

I. Paralysis of intra-uterine onset	Large cerebral defects; porencephaly. Defective development of pyramidal tracts. Agenesis corticalis (highest nerve elements involved).
II. Birth Palsies	Meningeal hemorrhage; rarely intracerebral hemorrhage. Later conditions: Meningo-encephalitis, chronica, sclerosis and cysts; partial atrophies.
III. Acute (acquired) Palsies	Hemorrhage (meningeal and rarely intra-cerebral). Thrombosis; syphilitic and marantic. Embolism: Terminal stages, atrophy, cysts, diffuse and lobar sclerosis. Chronic meningitis. Hydrocephalus (seldom the sole cause). Primary encephalitis: Polioencephalitis acuta (Strümpell).

Of this classification it should be said that brain defect or porencephaly, can occur as a result of birth or post-natal brain destruction. At least cases have been described in which porencephaly has been found post-mortem in individuals dying several years after trauma or the onset of a disease in a formerly healthy baby. Primary encephalitis is perhaps a good term though encephalitis unqualified would be better. The organisms causing the inflammation have, in most oases at least, caused some disease process elsewhere in the body before the cerebral involvement. Also encephalitis should be given a place much higher in the scale, among the causes for acquired infantile palsy it should be first or second instead of last.

Trauma is not mentioned in this classification. There may be a direct destruction of brain tissue sufficient to produce the symptoms of infantile cerebral palsy. More commonly, however, the trauma causes hemorrhage and the hemorrhage destroys the cerebral tissue.

It may be that under the headings meningeal hemorrhage and hemorrhage in II and III respectively, these traumatic destructions are included; they should, at any rate, be kept in mind.

Etiology.—Infantile cerebral palsies, as the remarks in the section on Classification show, do not constitute a single disease with a single etiology. Any factor acting in such a manner as to destroy brain tissue, and yet not kill, may be the cause of the syndrome.

Disease of the mother while carrying the child may predispose to, or cause brain destructions in the child. Any condition that causes a maternal anemia, may lead to failure of proper development in the fetus through improverished blood supply. Acute infections in the mother may be transmitted to the fetus and lodge in the brain with disastrous results to the infant though no outward signs are manifest until several weeks after birth. Chronic infections, syphilis and tuberculosis, may be transmitted and act in the same way.

Unfortunately in most of these conditions the etiologic agent is undeterminable from the scars found at autopsy.

The rôle of syphilis in the causation of infantile brain destruction is growing in importance just as the rôle of emotion of the mother is diminishing. Freud, in Nothnagel's System, in 1897, showed that Abercrombie,[15] Sachs and Peterson,[16] and Marie[17] all were unable to demonstrate syphilis in infantile cerebral paralysis with any degree of frequency. At present many fairly conservative men believe syphilis to be the cause of a large proportion of infantile cerebral paralyses. Erlenmeyer,[18] in the early nineties, was a radical believer in the causative rôle of lues, even going so far as to state his belief that many cases, supposedly due to an acute infection, were, in reality, due to a latent syphilis lit up by the acute process. When one considers, first, the predilection of the spirochete for the walls of the blood-vessels and the lesions produced therein, and second, the rôle that hemorrhage or thrombus plays in infantile cerebral paralysis, the idea of Erlenmeyer ceases to appear fantastic or even as a gross exaggeration.

It has taken what seems like a long time for the profession to accept syphilis as a causative agent in this group of syndromes.

The work of König[19] in 1898 shows a curious attitude towards the problem of etiology of infantile cerebral palsies. He divides the etiologic factors into predisposing and true causes. The predisposing are:

1. Nervous heredity—nervous or mental disease in the parents or grandparents.
2. Alcoholism in the father.
3. Tuberculosis in the parents.
4. Syphilis in the parents.
5. Psychic or somatic trauma to the mother during the pregnancy.
6. Intermarriage of blood relations.
7. First born.
8. Prematurity.
9. Illegitimacy.
10. Child weak from birth.

The true causes are:

1. Difficult birth or asphyxia at birth.
2. Head trauma.
3. Infectious diseases.

To put number 4, syphilis, in the same class as numbers 10, 9, 8, 7 or even 6, seems like clouding the real issues of the problem. The difficulty of determining the presence of syphilis in 1898 was much greater than it is now. This doubtless accounts for the lack of emphasis put upon it. The rightful position of syphilis is not established yet, but it surely deserves a much more prominent position than that assigned by König.

There are a few facts known about syphilis that apply to this problem. Syphilis in its second stage is a vascular disease; it attacks the walls of blood-vessels in all parts of the body. In these vessels it leads to two events, either of which can underlie infantile cerebral destruction, i.e., vascular mural thickening with attendant narrowing of the lumen, and, mural weakening which may lead to rupture and hemorrhage. These vascular changes begin within the first year of the infection, hence their effects may be looked for during the first year of infantile life.

It is also known that syphilis can exist for some time in the body without manifesting its presence until some serious vascular lesion of the central nervous system suddenly appears. Also, absolute dependence cannot be placed upon statements of parents as to the presence or absence of infection in themselves.

The number of cases of juvenile paresis and tabes is great enough to confirm us in the opinion that syphilis is inherited and does, in this exhausted form, often attack the central nervous system.

As to whether in its inherited form it produces infantile cerebral palsies with, or more rarely, without mental deterioration, or juvenile paresis, is largely a question of the age at which the cerebral changes begin to show themselves clinically. If early it is because of the involvement of a relatively large vessel with motor symptoms, then the picture will continue to be essentially vascular; if late the lesions are usually parenchymatous, or, clinically, paretic.

Savey,[20] in 1909, collected the cases of infantile cerebral hemiplegia in which syphilis was the etiological agent. At that time he collected over 30 cases from the literature. They included cases of hydro-encephalus (Hasse[21]), cases with associated blindness due to optic nerve atrophy or to keratitis and cases in which there was a transient paralysis within the first few months of life followed after a few more months by a permanent disability. These last cases show most clearly their endarteritic or syphilitic origin. The spasm or transient closing of the vessel with paralysis of short duration is what is so frequently seen in the syphilitic diseases of the adult brain. Hadden's[22] case is briefly as follows: History of lues in parents; infant suffered an eruption and coryza at two months, then a sudden right hemiplegia, improved on specific treatment. At three months convulsions and right hemiplegia. At 14 months infantile spastic hemiplegia and idiocy. Seibert[23] reports a case in which there was a left hemiplegia which cleared up with specific treatment, but returned within a month in a permanent form. Babonneix and Tixier[24] report a case in which the birth was normal. The baby talked at the usual time but did not walk until the nineteenth month. At three there was headache, then convulsions and a left hemiplegia. Parents' history was indicative of lues, though the mother's serum gave a negative Wassermann; the baby's serum gave a positive Wassermann. Pehu and Gardere[25] describe a case in which autopsy showed an acute encephalitis which was not to be distinguished from the ordinary non-specific encephalitis except by the demonstration of the spirochete.

Improved methods of detecting syphilis have secured its recognition in many cases where previously it was scarcely suspected. This has led to an appreciation of its importance as an inherited factor and the idea of inheritance from grandparents through apparently non-syphilitic parents has gained great credence. Quite recently such high authority as Gaucher[26] of Paris has insisted upon a recognition of the rôle of syphilis in the causation of idiopathic epilepsy.

There are means of demonstrating syphilis that are wanting in the acute infections. Therefore, syphilis will show an increasing ratio as a causative factor as the newer methods are applied. The number of cases in which emotional and physical trauma of the mother are blamed will show an accompanying decrease. It will not be proportionate nor will the decrease reach its true low level until means are devised and used for determining the occurrence of acute non-specific inflammations of the fetal brain.

The pregnant mother, in looking back in search of cause of her child's paralysis or chorea, may easily fail to recall an acute infection but will remember a terrifying experience. It is easily conceivable that microörganisms and the signs of acute inflammation can be destroyed and absorbed, grown around, if the acute process occurs to the embryo brain at a sufficiently early period. In fact this is one of the three hypotheses, namely, inflammation, thrombus, or hemorrhage upon which porencephaly can be explained, otherwise we must fall back on some such general terms as congenital malformation.

Premature birth has been frequently given as the cause of infantile cerebral paralysis. That the mere fact of prematurity cannot be the sole cause of the symptoms the numerous premature babies who show no signs of brain destruction sufficiently attest. The rôle of syphilis and tuberculosis (Wall[27]) especially the former, has long been recognized in the etiology of premature birth (Placher[28]). It seems much more probable that these chronic inflammatory processes inhibit development or cause destruction, than that the mere change from intra- to extra-uterine life brings about the morbid process.

Just how they act is largely hypothetical. The rapidity of intrauterine growth would suggest that the conditions for successful life are much better before than they are after birth. It may be that development of the syphilitic fetus within the mother can go on but that such progress can only be made within the mother. The cellular architectural arrangement of the brain cortex undergoes marked changes up to the time of birth. Marked changes in the body, of a developmental nature, need the best kind of support, call for great effort. Obviously such support and effort can scarcely be expected of an infant with narrowing of the cerebral blood-vessels, or with any other causes of nutritional impoverishment.

Since Flechsig[29] discovered the late myelinization of associational and pyramidal tracts, prematurity has been thought of as interfering with development through interference with this myelinization. It is quite possible, according to the work of Mellus,[30] that cellular development is retarded quite as much, and as importantly, as myelinization, and the spastic paraplegia which occurs more frequently in those premature babies may be of cortical, cellular origin instead of lower myelin or fiber origin.

Kehrer[31] collected 32 cases showing structural abnormalities of the skull of the newborn which he considered due to presence of amniotic adhesions. These adhesions occurred as a result of trauma to the pregnant uterus, dermoid cysts, retention cysts and other tumors. This source of disturbance to normal cerebral configuration is of interest as a rather unusual pathologic condition, but it is questionable if it occurs frequently enough to warrant a prominent place in the etiologic consideration of the disease.

In 1862 Little[4] published his classical articles showing the rôle assumed by birth trauma in the production of infantile cerebral palsies. But it was not until 23 years later that the real significance of this trauma was indicated when Sarah McNutt[5] showed that the essential immediate lesion was a meningeal hemorrhage.

There has been much discussion since 1885 as to what causes the hemorrhage. It is believed that in prolonged labor, when the head is fixed in the pelvis, a venous congestion takes place with marked over-distension of the sinuses. At the same time the molding of the head continues. If this has to be extreme, because of a small pelvis or a large head, then overriding of the parietal bones will occur and some of these over-distended veins will be torn. This theory is evolved partly to explain the preponderance of convexity hemorrhages. This action of the overlapping parietal bones is discounted by the fact that, as Kowitz[32] showed, many of these babies dead of meningeal hemorrhage, also had hemorrhages elsewhere, as in the pleura, the cardiac region, etc.

Precipitate labor has been blamed for causing infantile cerebral palsy also. The rapid passage of the head through the birth canal did not allow of time for molding and so hemorrhages were provoked from vessels to which sufficient time was denied to prevent over-distension.

Much discussion was waged about the part that forceps play in the causation of hemorrhage. The obstetricians naturally are anxious to believe that prolonged labor is more dangerous than the application of forceps. Statistics seem to bear them out in their contention. From the consideration of a very large number of mentally abnormal children Stein[33] concluded that instrumental labor was less dangerous than prolonged labor. He quotes various other authorities in confirmation of that opinion.

There is no doubt that a greatly prolonged labor is dangerous to the fetal head, but to advise simply an increased use of forceps seems unwise. Forceps in the hands of some men may be highly desirable but in the hands of others greatly to be feared.

The after-coming head in breech presentations and following version seems to favor hemorrhage (Holt[34]). This supports the belief that venous stasis and asphyxia are important factors in the production of hemorrhage.

The exact rôle played by syphilis in the causation of meningeal birth hemorrhage is not fully determined but unquestionably anything that will weaken the walls of the blood-vessels will increase their liability to hemorrhage. So a syphilitic child born through a small birth canal by a sick and weakened mother is especially liable to birth hemorrhage. Weye, quoted by Vogt,[35] says that he found syphilis in one-fourth of the children who died as a consequence of intracranial hemorrhage.

In cases in which the disease is acquired after birth, infection plays much the largest rôle. Measles, scarlet fever, whooping-cough, diphtheria, pneumonia, typhoid fever, mumps, and streptococcic disease of the tonsils are the frequent forerunners of an encephalitis which leaves behind it a case of infantile cerebral palsy.

Meningitis is one of the most important of the acute infections. Meningitis may occur in the course of any of the acute infectious diseases of childhood. It appears as a complication much as the bronchopneumonias do. In case death is not the outcome, it may produce cerebral spasticity either through cortical destruction due to an associated encephalitis, or it may set up a chronic hydrocephalus.

A variety of meningitis which in itself may cause cerebral spasticity is pachymeningitis hypertrophica. The statement that in itself it may produce cerebral spasticity or cerebral destruction should be qualified. The pressure may be enough to produce symptoms, just as symptoms are produced by the overlying hemorrhage or the cortical destruction, or by the action of the toxins generated in the meninges.

The acute infections of the gastro-intestinal tract may play an etiologic rôle in two different ways. The acute inflammation of the brain may occur during the acute stage of the intestinal infection, in which case it is caused by a bacterial embolus locating in the brain. Or the marasmus into which these children sink may occasion a thrombus or even a hemorrhage of the brain. Or again the enfeeblement of the child after a prolonged enterocolitis may predispose to any chance infection, or even to an easy spreading of the original infecting organism.

There is a further manner in which intestinal disease may play a rôle in the causation of brain disease. Spiller,[36] under the title, "Severe Jaundice in the Newborn Child a Cause of Spastic Cerebral Diplegia," describes four cases which showed jaundice during the first week of life. In three of the four cases a typical spastic diplegia resulted; in the fourth case (Case III) there was a hypotonia of the neck muscles. The author cites Pitfield's[37] four cases of icteric infants. In two of these cases the skull was opened; in one, at autopsy, 72 hours after birth, revealing a subdural hemorrhage; in another, at operation at five, disclosing adhesion of dura at cerebellum. He also refers to Schütte's[38] article in which a case of disease of the frontal lobes was accompanied by disease of the liver; and to Bostroem's[39] case of pseudosclerosis in which Bostroem ascribes the brain disease and the hepatic disease alike to an intestinal toxin. The progressive lenticular disease of Wilson belongs in the same category.

If, in the course of any acute infection, convulsions, somnolence, unconsciousness, or delirium, develop, one must be on the lookout for signs of paralysis, mental weakness, or other indications of brain destruction. The acute period may be with or without signs of meningeal irritation but usually there is at least slight rigidity of the neck, headache, and some cells in the spinal fluid.

Hemorrhage may occur in the infant but probably it never does unless some disease of the blood-vessels has weakened their walls to near the breaking point. The cases of Pitfield's in which subdural hemorrhage was found or suspected in the presence of jaundice, bears out the old idea that jaundice predisposed to hemorrhage. Now it is generally accepted that jaundice may be an evidence of blood destruction in the liver, hence the escape of the blood from the vessels would seem to be the primary process occasioned by some disease of the blood or of the containing vessels. Thrombosis of the cerebral vessels may occur either from disease of the vessel walls or in marasmus. It is most frequent in luetic vascular disease. The intima is compressed and then broken through by the specific infection of the vessel wall. The protruding roughened wall furnishes an ideal point from which an occluding thrombus can develop.

Trauma of the skull may be the cause of infantile cerebral paralysis, though its exact action is somewhat in doubt. It may act by destroying brain tissue or by causing hemorrhage; in these cases there is no question of its action. In some cases where trauma of a less destructive degree is blamed for the disease it is possible that the trauma has merely wakened a dormant syphilis, or has caused a locus minoris resistantiæ in which an acute inflammatory process has gained a foothold.

Fright has been given as a cause of infantile cerebral paralysis, but as more exact methods of examination are employed the necessity of bringing in fright as an etiologic factor lessens. The remarks relative to the rôle of emotion in the mother apply here as well.

Pathological Anatomy.—The initial lesion of cases of infantile cerebral palsy frequently cannot be determined from the findings at autopsy. The interim between the onset and death may be many years and all signs of the primary disease will have disappeared. The scar will remain but will give no more indication of its cause than a scar on the skin.

We know enough, however, to feel fairly safe in stating that the initial lesion in the vast majority of cases is either of vascular origin, a hemorrhage, thrombus, or embolus, or of inflammatory origin. In a sense the latter is a subhead of the former since the inflammatory agent is conveyed to the brain in the blood stream. The work of Marie and Jendrassik first made it clear that the sclerotic processes of Cotard and Strümpell were of vascular origin; that they spread out from the smaller arterioles and capillaries.

Cotard,[2] a pupil of Charcot, in 1868 classified the findings, the end results in infantile cerebral palsy as follows:

1. Yellow placques.
2. Cysts and cellular infiltration.
3. Complete failure of nerve tissue.
4. Lobular sclerosis.

It is simple enough to see that the yellow placques arise as vascular lesions. They are similar to the same condition found in the adult due to thrombosis and softening.

Likewise the cysts can be understood as arising from hemorrhage, embolism or thrombosis. Complete failure of nerve tissue or porencephaly, as it was called by Heschl in 1859, was attributed by him to disease of a blood-vessel in fetal life.

It is more difficult to attritubte to vascular disease the diffuse sclerosis. It may be, in many cases, that it is attributable to vascular disease only in so far as the vessels carry the inflammatory agent to the particular part of the brain.

To go back to the yellow placques. They are surface softenings due to closing of vessels entering the brain from the meninges. They may be small or they may be several centimeters in diameter. They do not differ from similar lesions in the adult.

The thrombus arising from vascular disease or embolus plugs the vessel, the tissue supplied by it degenerates, all the elements disappearing until only an amorphous unabsorbable yellowish-brown placque lies beneath a thickened pia with cerebrospinal fluid filling in the lost space. The color comes from blood pigment.

The location and extent of these placques are determined by the vessel affected. Vessels in the distribution of the sylvian artery are most frequently involved. These vessels, the cortical branches of the middle cerebral artery, are distributed to the tips of the temporal lobes, to all but the anterior fourth of the second frontal convolution, to the first and second temporal convolutions and to all but the superior third of the central and parietal convolutions.

The anterior cerebral supplies the tips of the frontal lobes and the posterior cerebral the tips of the occipital lobes.[40] The tips of the lobes may be bilaterally affected by trauma such as a fall in which the skull is not fractured but the force causes the brain to bruise against the skull. In such a case I found the tips of both temporal lobes converted into yellow placques extending back for about 4 cm. Lues was not to be excluded in this case; the fall may have predisposed to the location of luetic vascular disease.

The cysts are formed by thrombosis of deeper lying vessels. The walls are usually stained with blood pigment and from the walls the cellular infiltration spreads out into the neighboring brain substance. The cellular increase is of glial nature and paralleled by an atrophy of true parenchyma.

The cause of this thrombus is not infrequently indeterminable. In many cases it is luetic. If it is luetic there will be cellular infiltrations of the vessel walls and of the tissue of the immediate neighborhood. The elastic layer of the small arteries will show a characteristic splitting and reduplication. If special stains are used the spirochete may be demonstrable.

Porencephaly is usually a condition of fetal origin. Most authors have followed the example of Heschl in ascribing it to closure of a vessel in fetal life.

Some discussion has arisen as to whether the term is properly applied in cases except where the defect led to an opening into the lateral ventride. Heschl applied the term to cases in which there was a large defect of brain substance, regardless of its relation to the ventricle.

Porencephaly is frequently bilateral and more or less symmetrical, though as Kahlden[41] pointed out it may be of larger extent in one hemisphere and merely suggested or indicated in the other, or in the hemisphere in which there is no defect there may be only a well-marked anomaly of the gyri.

The relationship of cysts to porencephaly seems rather close, in some instances there is nothing to determine whether a given brain defect is to be called a cyst or porencephaly. If a cyst has only a narrow wall separating it from the ventricle and is covered by only a thin membrane, there is very little to distinguish it from the porencephaly defect which appears just the same except that it opens into the ventricle. The products of degeneration are present in the walls of the cysts in varying degree. In some long-standing cysts they are negligible, then the cyst wall will closely resemble the wall of the porencephaly defect.

In a few cases the wall of the defect has shown signs of cyst formation but for the most part no scarring or healing process is detectible in the walls. Its absence, or presence, is probably determined by the duration of the interval between onset and autopsy, in which absorption is taking place. The defect usually occurs in the area of distribution of the middle cerebral artery. If the lesion is of early fetal origin there is well-marked variation to be seen in the configuration of the gyri, but if it has occurred late, i.e., after the gyri have been established, no alteration is to be found.

Lobular sclerosis was at one time considered as a process peculiar to the brains of infants and its inflammatory origin was unsuspected. In 1885 Strümpell[10] published his article claiming that in the brains of children a disease occurred similar to or identical with poliomyelitis. Shortly thereafter Marie[11] put forth his views on the nature of the process; views which are accepted to the present day. He disputed Strümpell's idea that there was any specificity in the disease, or that it affected only a certain portion or tissue of the brain, or that it was caused by a specific microörganism.

Marie believed as we do now-a-days that acute inflammation could occur in the brain in the course of any of the acute infections, and that its exact location was largely accidental.

There is no doubt that the point and time of entrance of an infecting organism may escape notice and its presence in the body be suspected only with the occurrence of general signs of intoxication or local signs of inflammation.

In the infant, acute encephalitis is not nearly as fatal as it is in the adult. It is an acute inflammatory action. The inflamed organ swells. This it can do in the distensible skull of the infant, but not in the rigid skull of the adult. In the latter the swelling compresses the medullary centers and causes a respiratory paralysis.

This inflammatory process was shown by Jendrassik and Marie[3] to proceed from the smaller arterioles and capillaries. Hence, it is to be considered as a hematogenous infection; the primary focus may be anywhere in the body.

The sclerosis as seen in the late stages consists of an overgrowth of glia cells with corresponding destruction of ganglion cells. The typical perivascular infiltration gradually disappears as time goes on.

Macroscopically the brain is small in gross bulk. The loss of size may be circumscribed to one lobe or may spread throughout the area supplied by one of the three chief arterial supplies or an entire hemisphere may be involved; it is not uncommon to find it fairly symmetrical; it is rather more common in the area supplied by the middle cerebral artery than elsewhere. In some cases the atrophy may be so extreme as to lead to an uncovering of the insula or even the cerebellum.

The individual gyri are smaller than normal. They may be rather closely packed together as in the normal brain, or the sulci and fissures may be broadened and deepened with edematous overlying pia-arachnoid. In all these conditions an enlargement of the ventricles is the rule.

As has already been said, the pathologic anatomy of birth palsy depends upon meningeal hemorrhage. Kowitz,[32] in recording the post-mortem findings in 5998 children, 16.9 per cent. showed intracranial hemorrhage or its trace. He said it was seldom extradural. In 55.2 per cent. of cases it originated in the dura; in 23.2 per cent. in the arachnoid; in 11.7 per cent. the hemorrhage was in the ventricle and in 9.9 per cent. in the brain substance.

In most cases the hemorrhage has disappeared at the time of autopsy. The degree of absorption will depend upon the length of time which has elapsed between the time of the hemorrhage and death. I do not say birth, as there is some doubt in many cases as to the exact time of occurrence of the hemorrhage. Lindemann and von Marnholtz[42] cite two cases of meningeal hemorrhage in which it would be difficult to localize exactly the time of bleeding. In the first, their Case II, the etiologic factors are: Mother working hard, leaning over, just before delivery and labor occupying only 20 minutes. No asphyxia noted at birth. Convulsions began at six weeks. At six months typical generalized spasm increased to convulsive intensity by handling. At autopsy a large meningeal hemorrhage; all tracts of the cord diminished in size and an aplasia of the pyramidal tracts. These spinal cord findings speak for a very early onset of the process; they suggest the possibility of a prenatal onset. In the second case, their Case III, with no etiology at all, stiffness of the legs was observed at birth. At three weeks the baby had spasms. It made no progress towards voluntary movement, or speech. When examined there were nystagmus and convergent strabismus, bilateral facial paresis, legs spastic in extension and crossed, and arms flexed. At autopsy, death occurring at 15 months, a meningeal clot was found adherent to the dura, there were hemorrhagic cysts throughout the cortex and both crossed and the left anterior pyramidal tracts were lacking.

In speaking of hemorrhage as a cause of infantile cerebral palsy the authors say it may occur in the meninges or brain through trauma to the skull, before, during or after birth.

In the latter of these two cases death occurred at 15 months. No statement is made as to the possibility of there having been recurrent extravasations, so that we may conclude that absorption had not been completed during this period.

Pachymeningitis may be a cause of infantile cerebral palsy. As mentioned in the section on "Etiology" it may cause symptoms directly by pressure or indirectly by toxic action. The severity of brain symptoms in some lesser pachymeningitis lesions rather inclines one to the belief in the toxic theory. At any rate the evidence all seems to prove that the meningitis itself is due to the action of an inflammatory agent rather than the outcome of repeated hemorrhages. Schwartz[43] says that as long ago as 1856 Virchow said that the membrane preceded the hemorrhage. Also the work of Marie, Roussy and Laroche[44] seemed to show that the mere presence of blood under the meninges was insufficient to set up the hypertrophic inflammation. According to their experiments, repeated subdural injections of blood failed to set up a pachymeningitis.

Kowitz[32] found intracranial hemorrhage in 16.9 per cent. of autopsies on 5998 children, but he found pachymeningitis in only 3.9 per cent. In Schwartz's case, that of a five months old girl, there was a furunculosis at two weeks, at four months convulsions, vomiting, 20,600 leukocytes, hemorrhagic chorioretinitis, and the meningococcus was isolated from the spinal fluid. This seemed to show the inflammatory nature of the process.

Finkelstein[45] says cases of supposed rachitic malformation of the skull may, in reality, be cases of pachymeningitis. He considers the two important factors in the production of the pachymeningitis are repeated slight injuries and infection.

Grosz[46] reports a case of extra-uterine pregnancy in which pachymeningitis was ascribed to pressure on the skull during the extra-uterine gestation.

Hydrocephalus occurs frequently in infantile cerebral palsy. It may be of but slight degree and unilateral or of a very high degree and bilateral. In all cases in which there is a moderately extensive destruction of cerebral substance there is an enlargement of the lateral ventricle of the affected hemisphere. This is probably merely a compensatory process; the space within the skull cavity must be filled up, cerebrospinal fluid is the natural filler and cerebrospinal fluid is produced within the ventricle. Hence the ventricle will enlarge itself. This is perhaps not a true hydrocephalus yet it is difficult to draw a distinct line between cases of this sort and cases in which the ventricle is enlarged in an active rather than in a passive manner. Frazier[47] classifies the forms of hydrocephalus as follows: (1) Hydrocephalus non-absorptus. (2) Hydrocephalus obstructivus. (3) Hydrocephalus occultus. (4) Hydrocephalus hypersecretivus. The first variety is due to a cutting off of the arachnoid space by adhesions or is due to the presence of a toxin in the blood which prevents absorption. The second, as its name implies, is purely mechanical; either congenital defect or inflammatory adhesion has obstructed the channels of outflow from the ventricles. By the term occultus he designates the cases in which because the cranium undergoes no enlargement the condition remains hidden. In the fourth variety there is a hypersecretion of fluid by the choroid plexus owing probably to toxins in the circulating blood.

In all of these varieties it is the pressure of the ventricular fluid which brings about the ventricular enlargement. The choroid plexus is constantly secreting. In the first three varieties the fluid is not being reabsorbed. In the mild degrees the increased amount of fluid will interfere with normal development processes, in the more marked degrees it will cause active destruction or pressure atrophy.

Browning[48] gives as a factor in the production of over-secretion (the fourth variety of Frazier) pressure on the vein of Galen. He also ascribes the frequency of hydrocephalus in children to the fact that whereas the animal types of efferents or drainage, namely along the spinal nerves, exists only during fetal life, the Pacchionian bodies, though efferent structures in man, develop only after several years, in fact are not strongly developed at 20. Hence, there is only a very limited efferent or drainage system during the intervening years of childhood.

The ventricle, as above stated, may be but slightly enlarged or the greatly enlarged brain, up to twice normal size, may consist of a large thin-walled pouch. In the latter case the enlargement may be bilateral in which case the cortical wall is rarely so thin as in the unilateral cases. In the latter the more normal hemisphere may be completely covered over by the greatly distended pouch. The thin wall of the pouch shows marked alteration of structure, of both the white centrum ovale and the gray cortex. The cellular content of the latter may consist almost exclusively of glia in the marked long-standing cases. In the unilateral cases the contralateral cerebellar hemisphere may show a marked atrophy.

The condition of the pyramidal tracts in infantile cerebral palsy varies from apparent normality to complete atrophy. In the apparently normal cases slight variations may occur, the import of which is far from being understood. It is not known how much the spastic, paretic phenomena may depend upon these minor departures from the normal. For example, in Railton's[49] case of a three year old idiot with spastic diplegia the brain showed a slight depression over the central gyri with thickened adherent pia. The pyramidal tracts were intact, but the ganglion cells of the central cortex were diminished in number. In Rolly's[50] case the pyramids were declared to be normal but a generalized gliosis throughout the brain and spinal cord was described. Philippe and Cestan[51] describe four cases of infantile spastic palsies with normal pyramids. Mya and Levy,[52] cited by Miura, found a failure of the Nissl bodies in the cells of Betz; the pyramidal fibers were very fine and the myelin sheath was not normal. These microscopic changes were in a macroscopically normal brain and cord. Miura[13] cites the case of Spielmeyer, also, to show that the changes in the pyramidal system need not be the cause of the spastic palsy. Spielmeyer[53] found the cells of Betz and the pyramidal fibers normal, but found entire layers of the cortex destroyed: "Während ganze Schichten der Rinde zugrunde gegangen waren." He concluded that spastic paralysis might arise if the pyramidal system was isolated from its cortical connections. The effect of this isolation might be compared to the effect on the voice of the completely deaf.

Rhein[54] reported three cases of contracture and spasticity in which the pyramidal tracts showed only minor departure from the normal.

In one of the cases, the only case of cerebral palsy of infantile origin, he says there were no Betz cells found and the fibers of the pyramidal tract were very fine.

In going over this literature of the cases reported as having normal pyramidal tracts one is struck with the fact that in several the statement of the normal condition is frequently qualified by the addition that either the Betz cells are changed in number or staining characteristics, or else the pyramidal fibers are abnormally fine.

The pathologic condition of the basilar ganglia has not been sufficiently studied in infantile cerebral palsy to permit of a statement as to what their relation is to the clinical phenomena. Lesions restricted to the basilar ganglia are very rare in infantile cerebral palsy. Landouzy[55] described such a case in a woman dying at 32, who since two had had athetosis of the right arm. She had a bean-sized calcareous lesion in the left lenticular nucleus. Oppenheim and C. Vogt[56] described a condition of "marbling" of the basilar ganglia, that is, of the caudate nucleus and of the major portion of the lenticular nucleus. In this condition there was a disappearance of ganglion cells and a very marked diminution of fibers. This pathology was the apparent cause of a condition which Oppenheim described as pseudobulbar palsy. This was different from the usual pseudobulbar palsy, however, in that there was marked athetosis. The cortex, internal capsules, pyramidal tracts, the ansa lenticularis and optic thalmus were all normal. They cite the case of Anton[57] presenting a severe athetotic diplegia with a bilateral destruction of the posterior four-sevenths of the putamen.

In neither of those cases was the lesion due to vascular disease. In the light of Wilson's work on progressive lenticular degeneration they are probably to be considered as cases of selective toxemia of some variety with resultant localized bilateral destruction of brain tissue.

In a case of my own in which there were very violent choreic movements, so violent that the child had to be kept in a crib bed with high sides, there was a very obvious smallness of the basilar ganglia, which has not as yet been worked out.

Symptomatology.—Infantile cerebral palsies are usually seen after the establishment of the clinical picture, after the various symptoms have become permanent. The picture then is the result of a scar or healed lesion in the brain with its attendant secondary degenerations and irritations. So when infantile palsies are spoken of we ordinarily think of the terminal stage such as comes into the office of the orthopedist. But just as acute anterior poliomyelitis has an active inflammatory or destroying period so infantile cerebral palsies have an active destructive period.

This active or acute stage of the disease may occur during intrauterine life when, of course, it will entirely escape observation. The child may be born at term and appear perfectly normal. Even a careful examination may reveal no signs of disease, as reflexes in the newborn are always brisk, movement choreic, and strabismus not infrequent.

The baby may be described as a "cripple," or as a "stiff baby" from birth, but it is doubtful if the true condition is suspected at the time of birth except in the cases of very marked abnormality, such as deformities of the skull. The anencephalies and the marked porencephalies with attendant hemilateral or bilateral cranial depressions are recognized, but a lesser degree of porencephaly may be unattended by any marked cranial anomaly, and so escape detection. At any rate, the infant has passed through the acute period without the possibility of recognition.

The birth palsies may not be recognized until the delay in sitting, talking, or walking calls attention to the abnormality. On the other hand, in birth palsies convulsions may occur immediately, or within a few days after birth, thus calling attention to the presence of the disease process. These convulsions may mark the occurrence of meningeal hemorrhage. If the child is examined at this time there may be but little to mark a departure from the normal; or, the signs of intracranial disease may be well marked.

The meningeal hemorrhage may cause distinct signs of meningeal irritation with retracted stiffened neck, with fixed head, and anomalies of ocular movement especially nystagmus and strabismus. The increase in intracranial contents will occasion a change in the fontanelles. They may bulge strongly or may merely lose their characteristic compressibility and pulsation. The general musculature of the child may show a decided and symmetrical lack of tonus or the tonus of one side may be increased or decreased as compared with the tonus of the opposite side. Lumbar puncture at this time may reveal the presence of blood in the spinal fluid. Bloody fluid from fontanel puncture is especially strong evidence of hemorrhage.

If the hemorrhage is large, somnolence, slow pulse, or a rapid weak pulse, are the rule. If the hemorrhage is not large enough to compress the brain and inhibit vital functions and allows movements, a difference will be noted in the use of the two sides of the body. There may be a great deal of difficulty in getting the child to nurse.

There may be hemorrhages elsewhere in the body that show symptoms. Kowitz[32] found hemorrhages in the pleura in 35.6 per cent., of the cardiac region in 32.4 per cent., and in the thymus in 19 per cent., of 549 stillborn babies who died with intracranial hemorrhage.

Difficulty in getting respiration started is the rule in these cases. Probably many of the children born dead, blue babies, are blue because of an asphyxia which may be induced by the intracranial hemorrhage or, on the other hand, the asphyxia may act as one of the causes of the hemorrhage. When respiration is once started the cry is weak with less than the usual amount of facial grimace. The associated movements of arms and legs of the normal child are absent, in toto or hemilaterally.

The acute stage is frequently seen in the acquired forms. It may come on without warning, come as a late complication of measles or scarlet fever or other acute exanthema or may come on during tonsillitis or other inflammation of the respiratory, or gastro-intestinal tracts.

Usually the onset is sudden. Convulsions may usher in the other symptoms of fever, vomiting, rapid pulse and stupor or coma. During this period, unless rigidity is present, due to meningeal irritation, the child may be in a state of abolished tonus, hemilateral or bilateral. The rigidity when present is of the neck and is associated with generalized hyperesthesia. Already at this early period foot clonus or brisk knee-jerk may be present in a limb which otherwise seems quite flaccid.

In most cases, soon after the acute symptoms have passed off the rather characteristic increase in tonus manifests itself.

During the acute stage, in those cases due to inflammation of the brain, an optic neuritis of even an extreme degree may be present. Swelling of the nerve head is much less common in the cases of hemorrhage or thrombus.

If the lumbar fluid is examined it may be found to contain an increase of globulin; this is the most frequent abnormal finding. There may be an increase in cells which in the early stage are largely polymorphonuclears, or in the more severe cases there may be blood in the fluid. In these latter cases, however, the typical picture of infantile cerebral paralysis may not have a chance to develop as they represent the hemorrhagic encephalitides whose frequent outcome is death. This interpretation of the presence of blood is only to be made when the other evidences of an acute febrile process are also present; bloody spinal fluid is also found in cases of traumatic origin where the trauma has caused a meningeal hemorrhage.

The cases in which trauma is the cause may be considered as having an acute stage; the stage during which the hemorrhage is progressing and before a readjustment to the increase in cranial contents is accomplished. During this period there may be generalized convulsions or Jacksonian convulsions. The signs of increased intracranial pressure may be well marked, *i.e.*, somnolence, stertorous breathing, slow pulse, and vomiting, and in case the hemorrhage is frontal or in the region of the orbits, there may be some swelling of the optic disc. Involuntary discharge of bladder and bowel contents are usual.

Blood will be found in the spinal fluid in case the hemorrhage is large or has a chance to pass through the foramen magnum. Or instead of blood, hematagenous pigment may color the fluid any shade from that of a deep cherry to a light May-wine color.

After a period of hours or days the acute symptoms disappear. This is followed by a more or less prolonged stationary period of weeks or months, or even years. Then improvement begins and walking and some voluntary movement of the arm may be established. As a usual thing walking is delayed until the third, fourth, or even the sixth year. Eventually another relatively stationary stage is reached in which the interference with voluntary movements continues unchanged and mentality seems to progress at a retarded rate.

During all this time, after the acute stage, in the great majority of cases, increased tonus and lack of voluntary movement have gradually brought about conditions of contracture until finally the typical end-pictures of infantile cerebral palsy are developed. Naturally these end-pictures depend upon the region of the brain involved and the degree of its involvement. In two important respects infantile cerebral paralysis differs widely from the cerebral paralysis of adult life. In the first place interference with trophic impulses may result in failure of growth, and in the second place, involuntary movements may be of such a character and degree as to overshadow the paralysis.

The trophic disturbances are best seen in the hemiplegic form because in this form the contrast with the normal arm and leg makes the failure of development on the paralyzed side the more prominent. Evidences of interference with growth, however, are easily seen in other forms. In the diplegias the small size of the limbs and the failure, in some cases, of even the trunk to grow, is very striking. For example, one of a pair of twins with cerebral diplegia is much smaller, both in trunk and limbs than his twin.

The trophic disturbances are really more interferences with growth than trophic disturbances as we think of them in the adult; ulcers, gangrene, etc.

König[58] in his article on hypoplasia arrives at certain definite conclusions. He proposes the term hypoplasia instead of atrophy. Some of the more important of his conclusions are as follows: Hypoplasia can involve both the length and the circumference of the limb. He makes five divisions of the distribution of the hypoplasia: Of one-half of the body, the extremities and some portion of the trunk on one side, merely the extremities of one side, one extremity or a part thereof, and of the entire body and extremities. He agrees with former authors that an early onset favors hypoplasia though the latter is not dependent upon the age of onset and the duration of the disease, also that it can fail in congenital cases; that, though there is no constant relation to the severity of the symptoms, yet the most marked cases are seen in the severe spastic paralyses; that it may be the predominant, even the only symptom. His "pathologic certainties" are rather too sweeping to be accepted in toto. For instance, he says hypoplasis can occur with a normal cord; that it can occur with a brain macroscopically normal. On the other hand he states that hemihypoplasia indicates a small opposite cerebral hemisphere and that a hypoplasia may be lacking in disease of the entire post-central gyrus.

The involuntary movements, chorea and athetosis, vary in degree from those which will occasionally occur during an emotional period to those of such intensity as to dominate the picture. They may appear merely as one of the lesser symptoms of the disease or they may attain a degree of amplitude and force such as to seem to eliminate the applicability of the term cerebral paralysis.

In the section on "Classification," the forms, whose predominant symptom is paralysis, were divided into hemiplegias, diplegias, and paraplegias. This sequence is in the order of their frequence. In discussing the clinical pictures, the symptomatology of hemiplegia will be taken up first.

A few remarks concerning the etiology of the hemiplegic form should first be made. Hemiplegia occurs more frequently in the acquired or post-natal palsy than in the prenatal or the birth palsies. It may be caused by any of the three or four fundamental processes mentioned in the section on "Classification;" inflammation, hemorrhage or thrombus, trauma, or congenital maldevelopment.

Congenital maldevelopment can cause hemiplegia only in the sense that porencephaly is a form of congenital maldevelopment; the other forms are all too distinctly symmetrical to cause a one-sided clinical picture. It is rare, indeed, that porencephaly causes hemiplegia as it usually is symmetrical and bilateral.

The clinical picture of infantile cerebral hemiplegia depends greatly on whether the paralysis is accompanied by marked trophic change, strabismus, involuntary movements (chorea and athetosis) or idiocy.

Ordinarily, trophic changes are well marked. The paralyzed arm and leg are shorter and their volume decidedly less than that of the normal side. The diminished size gives to infantile hemiplegia an appearance which clearly differentiates it from the adult form.

The atrophic arm is usually held firmly against the side. The forearm is flexed to a right angle and is half pronated. The hand is slightly ulnar flexed, the fingers are usually extended in their proximal phalanyx but sharply flexed in the two distal phalanges. The thumb is flexed into the palm where it lies under the fingers. In standing the thigh is slightly flexed on the pelvis, is rotated a little inward and adducted so that the paralyzed knee stands in front of and above the unparalyzed knee. The lower leg is slightly flexed on the thigh. The foot is in the equino-varus position and the toes dorsally flexed to almost a right angle. The weight is borne on the distal ends of the metatarsals.

In the severe forms there may be practically no voluntary movement in the paralyzed limbs though it is rare indeed that paralysis is so extreme as to prevent walking. The described positions of the various segments of the paralyzed extremities are due to contractures and may be strongly maintained against active, or passive movement. In some cases there seems to be no contracture. The paralyzed extremities can be readily moved, but fall back into the habitual position as though they were fitted only to that particular attitude.

A large number of statistics seem to show that the right side is involved more frequently than the left, in about the ratio 4:3. It may be that this is, in some manner, related to the essential cause of right handedness in general. This right handedness may be due to the difference in the angles at which the carotids are given off from the aorta. The left carotid is slightly more in line with the blood stream. The left hemisphere may have a slight advantage over the right in the conditions of blood supply under which it works. And so vascular accident and emboli, sterile or septic, would be more common in the left hemisphere than in the right.

In infantile cerebral hemiplegia the face is practically always involved, though it may be generally stated that it is less seriously involved than the arm. It is most noteworthy that the involvement is general; the upper portion is involved as much as the lower; and emotional as well as voluntary function is inhibited. In these two ways the facial involvement differs from that seen in the adult. For this, and for other reasons, it may be deduced that habit of action plays a large rôle in determining the distribution and degree of inhibition of voluntary movement in cerebral hemiplegias. In the adult the upper face is *habitually* moving in connection with movements of the eyes and in a sense their movements may be classed as acquired reflexes. The muscles of the lower face on the other hand are used much more voluntarily, being concerned in that most voluntary of human functions, namely, speech.

When destruction occurs before these habitual or acquired reflexes have a chance to be set up then we have a more apparent involvement of the upper face.

As in the adult, the arm is usually more involved than the leg. It seems to me that in a general way this is to be expected from any cerebral lesion.

Activities of the hand are much more voluntary than those of the leg. We hear of men going to sleep on the march but it is difficult, indeed, to imagine a man's hand going on writing or playing a piano during his sleep. So when cerebral impulses are shut off from one side of the body it is quite natural that the more a part of the body is engaged in voluntary action the more its function is going to be cut down.

As a rule tendon reflexes on the paralyzed side are exaggerated. Foot clonus and Babinski are usually present. Contractures and involuntary movements may interfere with demonstration of the reflexes but in themselves they denote that which the increased reflexes also indicate, namely, increased tonus. Foot clonus may be demonstrated by putting a little pressure on the foot sole and then tapping the Achilles tendon with the percussion hammer. The ordinary method of producing foot clonus, such as is used in the adult, may fail entirely to elicit the sign.

In most cases the deep reflexes in the unparalyzed leg are also increased. They may even be exaggerated to a definitely pathologic degree. In a few cases the reflexes are normal; they may be diminished or even lost. The variation in the state of the reflexes has not been correlated with a sufficient number of careful autopsies. Souques[59] reported a case of infantile cerebral hemiplegia with lost reflexes in a patient who had had meningitis at two with optic atrophy. At about 10 there was an angina, suspected of being diphtheritic, with a sudden right-sided hemiplegia with involuntary movements and arrested development of the paralyzed limbs. The ankle-jerk on the paralyzed side was feeble; the wrist-jerk fair; the others abolished as were all the deep reflexes on the other side. The Babinski sign was positive on the paralyzed side, negative on the other side. There was no contracture, although the condition had persisted for 20 years. Souques considered the change (the failure) of the reflexes as being due to the meningitis. He compared the condition to that of tabes dorsalis.

König's[58] report of the reflexes in 27 spastic hemiplegias was as follows: In 21 all jerks were increased (I suppose by *all* he referred merely to the paralyzed side); in three cases they were increased in only the upper or lower extremity; in two they were normal and in one diminished. König makes no comment on these figures. It may be that in many of these cases of diminished reflexes syphilis is the causative agent and a meningitis is present as in Souques' case.

Disturbances of voluntary movement vary from complete loss of motion to mere clumsiness. This variation in degree depends upon more than one factor, though perhaps most upon the amount or degree of involvement of the pyramidal tract. As an example of the milder form of disturbance, I would cite the case of E. J., age 20 years. Birth was difficult. From the information obtainable I believe a version was done. Artificial means had to be resorted to to start breathing. Three days after birth he had a "spinal disease," during the course of which his left eye turned in. He was a "stiff baby," didn't walk until he was three years old.

Now he presents an internal strabismus of the left eye and a well-marked facial asymmetry; the left face is smaller than the right. He is *left handed*. The right hand is distinctly clumsy; he cannot rapidly touch the tip of one finger after the other to the thumb tip. He hops quite well on the left foot. When he attempts to hop on the right foot he flaps his arms in a very clumsy manner and as he reaches the end of the room and starts to turn he falls against the wall. The deep reflexes are all very brisk, those on the right being more active than those on the left. The right abdominal and cremasteric are diminished. No sensory disturbance; distinct feeble mindedness.

The paralyzed left sixth, the diminished function of the left seventh combined with the clumsiness of the right arm and leg localizes the lesion in the upper medulla and lower pons on the left side. The lesion may be associated with others in the cerebrum to account for the enfeebled mental state; on the other hand, the enfeeblement was not marked and there may have been merely a single lesion.

Cerebral Diplegia and Paraplegia.—The cases of cerebral rigidity constitute a very interesting class of disease pictures. They were classified by Rosenthal[60] in 1892 as follows: (1) Generalized rigidity. (2) Paraplegic rigidity. (3) Bilateral spastic hemiplegia. (4) Bilateral athetosis. (5) Choreiform diplegia. (6) Atypical forms. In 1897, Freud[12] classified them much in the same way merely gathering 4 and 5 together as generalized chorea and athetosis and omitting number 6.

The etiology is the same as in the other forms of infantile cerebral palsies. Different authors hold different views as to the more concrete facts concerning the etiologic factors. Collier[61] states that these forms are for the most part congenitally determined. Freud emphasized the injuries due to forceps; he stated that one-sixth of all diplegias occur in premature birth, but did not, on that account, ascribe the disease to the prematurity, but rather the two to some other cause such as lues. Little[4] believed that the difficulties of birth accounted for a large number of these cases. Ganghofner[62] gave no definite cause in 42 per cent.; prenatal origin 31 per cent.; post-natal 16 per cent., and birth trauma only 9 per cent. König[63] minimized the rôle of birth trauma; of 260 cases of idiocy, difficult birth with or without asphyxia was present in only 39. In five forceps were used; in 17 no other factor could be held accountable. If we remember the varied indirect and direct causes König cited as possible etiologic factors we realize how industriously he sought to distribute the responsibility. Vogt[35] stated that these varieties of cerebral palsy in infants occur for the most part as the result of birth trauma.

Of this much I think we can be sure, the condition can occur as the result of prenatal and birth conditions, also that accidents can more easily escape detection at these periods than during extra-uterine life so that in the cases in which the period of onset is not determinable the probability is in favor of the two earlier periods. We also know that the fetus may be born with a central nervous system apparently intact and yet infected with syphilis and this infection can produce the destructions and the symptoms of infantile cerebral paralysis. So much for the period of onset. As to the etiologic agent it may be said that anything that can cause brain destruction and yet not kill must be considered.

Infection is the most common cause. If we included under this heading the cases in which the infection is confined to the blood-vessel walls we would have a large majority of the causes covered. The other important cause is trauma, trauma to the maternal gravid uterus or to the head of the child before, during, or after birth. It is to this class of cases that the term *Little's disease* especially applies.

The clinical picture is varied. The child may be totally incapacitated or may be but slightly affected. In the former class are the diplegias, who lie or sit, if they are able, in the position in which they are placed. The legs may be drawn up to the thighs, and the thighs on the pelvis. There is a certain fixity of the lower extremities which though due to contracted muscles appears to be partly due to a gradually acquired deformity of the joints. The feet seem to resemble hands more than they do in the adult. There may or may not be a good growth of hair at puberty about the genitals, which in their turn may or may not be well developed.

The trunk may appear fairly well developed, at least when compared to the extremities. The arms frequently look less affected than the legs, yet when it comes to a matter of usefulness they may be no better. They may be contractured; pressed against the sides, forearms pronated and flexed to a right angle or more, wrists partially flexed, fingers in a fist, or in a position resembling the main accoucheur. The head may be somewhat retracted or more commonly thrust forward. The face gives the appearance of a mask due to a rigidity or fixity of expression. There may or may not be a strabismus. Voluntary motion may be restricted to movements about the mouth concerned with eating and chewing. These children may never be able to talk. Of a slightly lesser degree are those who can follow one with their eyes; can speak, though in a very low or weak voice; and may even be able to perform a few voluntary acts such as to make purposeful movements of the index finger or thumb. In some cases the fixed contractures in the arms may give way to involuntary movements. Then there is a combination of spastic contracture and chorea though it has seemed to me that in these cases the legs are never as spastic as they are in the purer types.

In the cases in which walking is possible the child walks on his toes, the feet are slightly turned in, the ankles fixed, the knees slightly flexed and rubbing against each other, the thighs move as a piece with the pelvis which seems to move on the base of the leg that is still. The arms are thrown about in an incoördinate manner as though through excessive "mitbewegungen" or associated movement. The face may be contorted with grimaces which include the tongue. It seems as though the entire stock of volitional impulse resident in the central nervous system was set loose, but was working without direction or control.

As before mentioned speech may be very weak. It may be produced with a great deal of associated movement including not only the face but the hands and arms, even the legs and trunk as well. An interesting movement associated with speech is that of the muscles of emotional expression such as to produce the picture of impending weeping whenever speech is attempted. As seen in one patient this was much more marked whenever anything was done to stimulate emotional expression. Articulation may be indistinct as well as feeble and associated with this speech difficulty swallowing may be much interfered with. In one case this dysphagia became so much worse when the patient was disturbed that there was great difficulty in feeding him under any but the quietest of conditions.

Strabismus is common among these diplegic cases. In some cases it is doubtless due, in part, to abnormalities of the eye-balls with visual disturbances. In the remainder of the cases it is due either to the involvement of higher centers for coördinated movement or of lower centers involving some of the oculomotor muscles.

There is a peculiar motor phenomenon not infrequently seen in these patients which closely simulates mental torticollis. Neurath[64] speaks of a similar condition in a hemiplegic in which case he ascribes it to a spasm or increased tonus of the deep muscles of the neck on the normal side. I have observed it in a diplegic imbecile in whom it was much more like the mental torticollis of the adult than like an organic spasmodic phenomenon.

Mental enfeeblement is the rule in these cases. It is necessary in many cases to search for proof of this. In the first place the family may think the patient unusually smart because he remembers dates with great exactness. They lose sight of the fact that because he is confined to very limited surroundings with markedly limited experiences, there is little to cloud or distract memory. In the second place these patients may get along fairly well if not hurried or pushed; they may use their limited stock of information well enough to make special tests necessary to demonstrate the enfeeblement. On the other hand it is in this class of patients that are found many of the idiots and low-grade inbeciles of the institutions.

In a few cases there are pupillary disturbances suggestive of syphilis or fundus disturbances such as neuroretinitis. I have seen medullated nerve fibers, peculiar changes in shape of the physiologic cup, or unusual arrangement of vessels of the disc often enough to lead to the suspicion that these anomalies were in some way related to the original process. Disturbances of voluntary motion of the face are the rule in these cases. There doubtless are cases in which voluntary motion is unaffected. These are mostly the cases of paraplegia. However, even in these cases movements of the face may give a peculiar impression as of rigidity, incoördination or exaggerated associated movements. The facial involvement is more marked in the lower distribution than in the upper. It is safe to say, however, that disturbances of voluntary movement of the upper face are more common in the infantile cerebral palsies than in the adult.

I have purposely avoided the word "paralysis" and spoken rather of disturbances of voluntary movement with reference to the face. Rigidity is perhaps the best term to express that which is present in the majority of cases. This is seen in a certain restriction of movements of the lips and lower jaw in speech and eating. It is also seen in expressions of emotion giving rise to the spasmodic type of laughing and crying. Various choreic movements are also seen which sometime resemble the facial tics, those, at least, that may be spoken of as grimacing tics.

The degree of involvement of voluntary motion in the muscles of the limbs and body may vary all the way from slight clumsiness and rigidity to complete paralysis, such as is seen in the flaccid idiot. In most cases there is a paralysis of certain muscle groups associated with either spasticity or involuntary movements elsewhere. As stated above the spasticity and involuntary movements can interfere as completely with voluntary efforts as a complete paralysis but where they are present the amount of brain destruction is usually less and as a rule there will be response from some part of the body to volition. The legs may be completely paralyzed, the arms very weak and too choreic to permit their being used voluntarily and yet the muscles about the mouth or of the tongue may be taught to obey desire.

Dejerine[65] has reported two cases of spastic diplegia due to lesion in the upper cervical cord. In Case I a sclerotic patch, considered by Dejerine as luetic, occupied the center of the cord at the C_3 level. In Case II, which was reported by Dejerine in 1897[66] and is here reviewed, the lesion is at C_2. The condition in both cases is that of a spastic quadriple gia with intact intelligence, no involvement of the face and no epilepsy. There is no failure of growth of the extremities such as is seen in most cerebral cases. Dejerine states that when a high degree of spasticity is found in the arms, as well as the legs, with no disturbance of intelligence of facial musculature and no epilepsy, that one may venture the diagnosis of infantile diplegia due to lesion in the cord. In emphasizing the importance of the presence of epilepsy as a diagnostic factor in localizing the lesion in the brain he cites a very interesting case. It is that of a man of 79, of good culture and intelligence. The face, speech, and arms were normal. Epileptic attacks had occurred on an average of once a month since infancy. At the autopsy, porencephalic defects extending into the lateral ventricles were found in both hemispheres. In the right it lay at the junction of the lower and middle thirds of the fissure of Rolando; in the left at the junction of the middle and upper thirds of Rolando.

Oppenheim and Vogt[67] described a condition of "marbling" of the basilar ganglia, the caudate nucleus, putamen and globus pallidus, occurring in a mother and daughter and producing a condition spoken of as pseudobulbar paralysis. In both cases there was spasm of all four extremities with pseudocontractures, choreo-athetotic movements, and mental enfeeblement. The term pseudobulbar was applied because of the marked involvement of the facial muscles and those used in speech, eating and swallowing. The optic thalami, the internal capsules and the pyramidal tracts were normal.

In the paraplegic form there may be no evidence of involvement of the arms and face. For example: A patient of mine rode about on a little platform on which his legs were folded and on which he sat. He propelled himself about quite rapidly and was quite agile in pulling himself up into a chair. Reflexes were quite brisk in the arms and the face gave a peculiar rigid monkey-like appearance. At the age of 27 he had an epileptic attack thus bearing out Dejerine's statement of the importance of epilepsy as a factor in the differentiation of cerebral from spinal paraplegia. Premature birth is given as the most important cause of the paraplegic form by Oppenheim. Since myelinization of the pyramidal tracts is complete only at birth it is supposed that this incomplete myelinization prevents the complete development of the pyramidal tracts in cases of premature birth. For this reason cortical impulses cannot travel down to spinal centers. Van Gehuchten gave additional strength to this theory by stating that pyramidal fibers themselves extended only to the lower medulla by the eighth month. Collier[61] denies that prematurity bears any real causal relation. He speaks rather of maternal pathology, such as syphilis and acute infections. He also stresses breech presentations as favoring birth hemorrhage at the upper end of the fissure of Rolando. It is a seductive idea that because of the after-coming head stasis will be most marked in the vertex and one hemorrhage could destroy both leg centers, because of their juxtiposition. Conclusive, substantiative evidence from autopsy material is wanting.

Lesser Symptoms.—Strabismus and other abnormalities of movement of the eyes are very common in infantile cerebral palsies. They are especially common in the diplegias and paraplegias. Where a typical abducens paralysis or oculomotor paralysis exists, especially if there is some disturbance of movement of the arm and leg of the opposite side, a lesion of the mid-brain can be assumed. Such a case has already been spoken of above. Nystagmoid movements: Bilateral, involuntary, rapid, oscillations are frequently seen in the diplegias and are probably due to involvement of the cortical centers for movements of the eyes. Collier considers them as evidence of a spastic condition of the extrinsic eye muscles. Disturbances of vision, because of the mental state of the patient, are demonstrated only with difficulty. Attempts to make the child blink may reveal blindness, hemianopic or total, though the latter has usually been discovered by the parents through long observation of the infant. In some instances, however, it is impossible to determine whether blindness exists or not. When hemianopsia is present it is due to lesions involving the optic tracts in the occipital or parieto-temporal lobes, even as it is in the adult. Vogt[35] cites a case of attacks of hemianopsia and headaches in which autopsy showed a destructive lesion extending from the central to the occipital lobe. The position of the head may suggest the presence of the hemianopsia being turned towards the blind side.

Atrophies of the optic nerve, neuroretinitis, cataract, and keratitis are seen and should arouse a suspicion of syphilis as the causative factor. Disturbances of pupillary reaction may have a similar significance.

Disturbances of speech are of two varieties; those which are due to faulty enunciation; these are very common, and those due to aphasia, which are very uncommon.

The enunciation may be feeble, lisping, stammering, or it may be guttural. It is usually in the diplegics and paraplegics that the more marked disturbances are found. The slow, feeble speech is analagous to the slow, weak, voluntary movements elsewhere. The guttural speech may be very similar to that of the adult pseudobulbar paralytic in its nasal, explosive, throaty character. These speech disturbances are frequently associated with difficulties in swallowing.

Aphasia is a very uncommon symptom in infantile cerebral palsy. When it is found, in the acquired cases, it usually lasts only a short time. The ability of the cortex of one hemisphere to compensate for failure of the opposite, is much greater in the infant than in the adult.

Disturbances of sensation are rarely demonstrable. In some cases a blunting of pain sense may be apparent when it is possible to compare response on one side of the body to that of the opposite. In only one case did König demonstrate a hemi-hypalgesia.

Increased reflex jerks are the rule in infantile cerebral palsies, but many exceptions must be made to this rule. König's[68] figures are interesting in this respect. As stated on page 59, of 27 spastic hemiplegics he found all the reflexes increased in 21; increased in *only* the upper or lower in three; normal in two; and diminished in one. In one flaccid hemiplegic, on the other hand he found increased reflexes. In 12 paraplegics he found eight spastic with increased reflexes; two flaccid with increased reflexes; and one flaccid with normal reflexes. In 35 diplegics he found 19 spastic with all reflexes increased in 11; diminished in legs in one; not demonstrable (because of increased tonus) in two; normal in one. In the remaining 16 cases the reflexes were present or failed in the uppers; were increased or normal in the lowers.

The Babinski sign is present in a large number of cases and is especially constant in the cases where the pyramids are involved subcortically. It is not safe, however, to say that in every case where the Babinski sign is present that degeneration of the pyramids will be found.

There does not seem to be a constant relation between the condition of the deep and superficial reflexes. In many cases in which foot clonus and Babinski are present, cremasteric and abdominal will show no change. In the majority of cases, however, the superficial reflexes are diminished on the side of the body on which the deep reflexes are increased; in the para- and diplegias they may fail entirely.

Involuntary Movements.—The range of involuntary movements in infantile cerebral palsy extends from intention tremor, such as seen in multiple sclerosis, to the most violent jerking of an extremity or of the trunk. Efforts have been made to differentiate the various types localizing each in a particular part of the central nervous system.

Though ataxia cannot rightly be classed as an involuntary movement, yet it is an involuntary departure from purposeful movement and as such has been included with tremors, chorea, and athetosis, under this heading.

The form of ataxia seen in locomotor ataxia is observed but rarely in a pure form; it is more frequently associated with spasticity. It is due to a disturbance of those tracts and centers which are engaged in transmitting sensory impulses to the brain, or, more correctly to the motor cortex, where these impulses act as sources of information to direct the elaboration of cerebral motor impulses. Perhaps the most important of the centers of sensory function is the optic thalamus. As early as 1883 Demange[69] showed the importance of lesions of the thalamus and its immediate environs in the production of ataxia. The thalamus contains the end stations and synapses of the two tracts whose integrity is probably necessary for perfect coördination; the spino-thalamic and the cerebello-rubro-thalamo-cortical tracts. Hence lesions of the thalamus can cause a high degree of ataxia.

An element of ataxia is also introduced into the disease picture by superficial diffuse destructions of the cortex, such as is commonly seen following meningeal hemorrhage. If this destruction extends to the post-central gyrus it will involve the cortical representations of the sensory systems.

Intention tremors are common in infantile cerebral paralysis. Tremors are expressions of spasticity according to Hughlings Jackson, whom Wilson quotes to the effect that tremor is rigidity spread out thin and rigidity is tremor run together. Wilson states that lesion of the pyramidal tract alone will not cause tremor, also that this tract must not be too severely damaged or else no tremor will appear. In lenticular degeneration, in pure cases of which there is no involvement of the pyramidal tracts, tremor is a characteristic symptom. Hence lesions of the lenticular nucleus may be the cause of the tremors seen in infantile cerebral palsy. This is stating where tremors are caused not where they must be.

However, since it is believed that there is no direct connection of the lenticular nucleus with the cortex the presence of tremor in infantile cerebral palsy is rather in favor of a lesion of the lenticular nucleus or its subjacent structures. Wilson states that the tremor is caused by lesion of the lenticulorubro-spinal system. This acts to remove the steadying influence on the anterior horn cells.

The more important, or at least the more spectacular, forms of involuntary movement are athetosis and chorea. Lewandowski and von Monakow strongly protest against considering the two as varieties of the same entity, as Lewandowski states other modern writers do. There is no question but that the two occur together in cases of infantile cerebral palsy.

Athetoid movements are slow and stereotyped. They may occur while the rest of the musculature is at rest, they may be brought out only when voluntary effort is being made elsewhere, or they may be controlled by voluntary effort. For example: A six year old child, who was born at seven months and had never walked, could keep his left arm still when he was sitting only by grasping some object that was about shoulder high and far enough away so that he could reach it with extended forearm. The athetosis was started by his attempt to rise to a sitting position.

In connection with voluntary control of athetosis it is interesting to note that König states that involuntary movements are more frequent in idiots than in more normal children. Lewandowski considers athetosis as closely associated to rigidity. It is to be noted that Wilson states on page 475, Brain, 1911, in his article on lenticular degeneration that athetosis is in most cases associated with hypotonia. I have not found this association in infantile cerebral palsy, nor in the few cases of athetosis I have seen in the adult.

Choreiform movements are very common in infantile cerebral palsy. They show as sudden involuntary jerks of various muscles which may be but slight and cause no change in position, but which usually are complex with several muscles involved in such a way as to cause more or less violent movements of head, limbs or trunk. They are very rarely present in sleep. They may be practically continuous during the waking hours but the more common condition is for them to appear more as associated movements when voluntary movement is attempted, or, as evidence of emotion. A child who is lying perfectly quiet may be thrown into violent chorea when spoken to. The movements may be of rather small amplitude or they may be large throwing movements. The athetoid movements are usually considered as involving especially the hands, fingers, feet and toes; chorea of the entire extremities; Lewandowski states that one of the differences between athetosis and chorea is that the athetoid movements are always the same in the afflicted individual whereas in chorea now this and now that muscle group is struck in apparently a haphazard fashion. This may be true, but it is also true that there are certain repetitions in the choreiform movements that are very near copies of each other.

Athetosis and chorea may be considered, as Freud does, as evidences of less severe involvement than paralysis or rather as steps in improvement from the more severe paralysis. Many cases have been published in which as the paralyzed limbs begin to improve athetosis or chorea appear. According to the teaching of Wilson we would say rather that with restoration of function of the pyramidal tracts athetosis and chorea are given a chance to manifest themselves. He would lead us to think that they are due not to lesions of the pyramidal tract, but rather to lesions of the cerebello-rubro-thalamo-cortical system. Since the function of this system is supposedly to provide proper working conditions for the cortico-spinal or pyramidal tract then lesions of this system can only be shown by the patient if the pyramidal tract can function sufficiently so as to produce movement of a cortical type.

Lewandowski states that with the subsidence of a choreic movement the limb relaxes into flaccidity. He makes no definite statement as to the reflexes. Wilson says that loss of reflexes in chorea represents a complication and does not belong to the essential disease picture.

Associated movements are very common in infantile cerebral palsy. They are difficult to distinguish from choreiform movements in some cases. They appear to be the result of two factors. In order to make a voluntary movement an extra effort must be made. This superabundance of stimulus overflows into adjacent or homologous areas of the body. Some of the more common associated movements are those of the face. For example: Attempts to wrinkle the brows have frequently associated with them, crossing or upward movements of the eyes; attempts to close or open one fist will cause a like movement of the opposite. Similar movements are much less common in the legs than in the arms and face.

Idiocy and Epilepsy.—These two morbid conditions are considered together, as both are frequent sequelæ of infantile cerebral palsy and because of the indefiniteness of the relation of the lesion to the particular symptom in both.

There has been a strong tendency to ascribe all cases of so-called idiopathic epilepsy to infantile cerebral lesions and it is perfectly possible that the same close relationship exists between idiocy and early brain destruction.

In the articles in reference texts such as Freud's in Nothnagel, and Vogt in Lewandowski, the emphasis is all on the part that infantile cerebral palsies play in epilepsy. It seems to me that their action in producing mental inferiority is fully as important, in fact, perhaps more important. The proof of the presence of epilepsy is much easier than that of moderate degrees of mental deficiency. In fact it may well be said that there are no definite ways in which we can accurately measure the higher grades of imbecility. Perhaps the only way that they can be measured is by the degree of success with which these individuals meet the problems of life. Considered from this point of view, namely, how the individual enlarges his horizon, keeps his poise and succeeds in competition, then many would have to be accounted as deficient. It may be said that this applies as much to those in whom there is no suspicion of infantile cerebral palsy as to those who have been afflicted. That may be, but in order to get an intelligent idea of the mental symptomatology of infantile cerebral palsy it seems to me necessary to assume that any brain destruction is almost certain to cripple the organ's ability to function to its fullest extent. It is only when it can so function that there is full mental power. If any portion of the infantile brain is destroyed there is just that much less to record impression, elaborate reactions, and express the results. It seems highly improbable that we can expect the same results from a smaller, that we can from a larger mechanism.

The degree of mental enfeeblement can vary from that which might be extremely difficult or even impossible to prove to the most hopeless idiocy. Ordinarily these idiots show some disturbance of voluntary movement. The amount of this disturbance will depend upon the direct or indirect involvement of the motor cortex. By indirect is meant those cases of pyramidal isolation or intracortical paralysis of Spielmeyer as well as those cases with lesions in the course of the tracts of great importance for proper pyramidal function such as the cerebello-rubro-thalamo-cortical tract. The disturbances of movement may be only such as that present in a case already cited in which there was a clumsiness, lack of clean cut action, in the right arm and leg of a left-handed boy in whom there was a paralysis of the right sixth cranial nerve and a history of difficulty, "spinal disease," soon after birth.

Epilepsy is often spoken of as inevitably leading to mental enfeeblement. This is not always true. Epilepsy may be present for many years without obvious mental deterioration. If epilepsy is due, in the particular instance, to infantile cerebral lesions of the sclerotic type then with progressive cerebral destruction there will be progressive mental deterioration. Epilepsy is less of an index to progressive brain destruction than mental enfeeblement; it is more the mere signboard that indicates the probable presence of a lesion in the child's brain. The lesion may be in any part of the cortex. It has been said that only Jacksonian fits must originate in the motor cortex; that ordinary general convulsions may originate in any part of the cortex. It may be that some of the distinct visual auræ described in ordinary epilepsy are merely evidence of the location of the epileptogenic lesion in the visual cortex; a lesion that has persisted since early childhood.

Treatment.—Preventive measures are most important in dealing with the problems presented by this collection of syndromes. Pregnancy should be prevented in the epileptics, the microcephalics and in the sickly anemic women. Syphilis should be most studiously ascertained and rigorously treated before pregnancy is permitted. This rigorous treatment is as necessary in the fathers as in the mothers. From numerous histories it seems evident that good specific therapy will do splendid service in protecting the ovum and fetus. It is not uncommon to find a healthy child coming from a syphilitic mother, who has had miscarriages and dead children, if conception and gestation take place during a period of intensive, or even moderate, therapy.

Tuberculous parents should have children only under the most favorable circumstances. If the mother is actively tuberculous she must live under the best possible hygienic conditions.

The sudden appearance of albuminuria, slight thyroid enlargement, headache, increased blood pressure, with nausea or vomiting should immediately raise the suggestion that labor may have to be induced. If syphilis can be excluded these symptoms often are ascribable to an acute infection from some more or less hidden focus. These symptoms of spreading of, and reaction to, the acute infection on the part of the maternal organism show that the local process has broken bounds and may affect the foetal as well as the maternal organism. It would be interesting if all cases of adherent placentæ could be accurately traced, both, back to their possible source from some acute maternal infection and forward to see in what percentage of cases neurologic and psychic abnormalities develop in the child.

In the avoidance of birth trauma the forceps should be used only by the skillful, and even in their hands less, and Cesarean section more frequently resorted to where definite information is obtainable of pelvic deformities and deficiencies. It goes without saying that only the skillful surgeon should attempt this operation, as much depends upon the rapidity with which the delivery is accomplished. Cerebral anemia during the birth of the child should be studiously avoided though we have as yet no certain information that in itself it is a very dangerous condition if it lasts only a few seconds.

The immediate recognition and treatment of meningeal hemorrhage in the newborn as well as in the older child is a most important prophylactic measure. Sloan[70] states that the mortality in untreated birth hemorrhage is 75 per cent. within a few days of birth, while the majority of the surviving 25 per cent. suffers from nervous disease. He cites Paul to the effect that in 50 per cent. of births through a contracted pelvis retinal hemorrhages are to be found, but that they are also present in 40 per cent. of protracted and in 20 per cent. of spontaneous labors. Hence, merely their presence is not diagnostic of intracranial hemorrhage, or at least of hemorrhage large enough to cause definite defects. In attempting to localize the meningeal hemorrhage Sloan makes the following suggestions. Restlessness and crying are more indicative of supra- than of infra-tentorial hemorrhage. Respiration is affected sooner in infra-than in supra-tentorial bleeding, (pressure upon the medullary, respiratory centers can be occasioned by less blood in the restricted infra-tentorial space).

The most reliable signs of hemorrhage indicating the necessity for interference are bulging and loss of pulsation of the fontanel, convulsions, small pupils later becoming dilated, twitching of the face and arm. When these signs are present a lumbar puncture should be done with a small platinum needle. Blood may be present in the spinal fluid. It is probable that blood pigment is more frequently found than unchanged blood, as, for instance, in the case of Lippmann.[71] This was an infant born to a primipara with a slightly contracted pelvis whose pains failed after 13½ hours. Low forceps were applied, a 5½ lb. child produced with no sign of trauma. At 48 hours the infant began to have convulsions, several in a day, was restless, irritable and cyanosis gradually developed. The anterior fontanel bulged tensely and at 76 hours a lumbar puncture revealed a spinal fluid with a slight yellow tinge. Human blood serum was injected subcutaneously and Lippmann[71] considered a cure and a prevention established. The procedure of Welch was recommended by Lippmann in the injection of human serum for spontaneous hemorrhage, namely, 30 c.c. are injected every six hours. The treatment may be continued for a week if necessary. It would seem that the withdrawal of spinal fluid, in this case, allowed of recovery of central nervous tissue; at least it allowed of a resumption of function. To say with Lippmann that because of this withdrawal that all danger of secondary contractures, spasticities, possibly epilepsy and idiocy, has vanished is going a bit far.

In the treatment of the end stages of infantile cerebral palsies we must turn to surgery for most of the improvement. The forms of the disease in which most can be hoped for are those in which voluntary movement is present, but is interfered with by spasticity and contractures. Various measures have been advocated for relief from these obstacles. Tenotomies will relieve contractures to a degree if the limb is placed in a more satisfactory position and kept for two to six weeks in a cast. Contractures of the hamstrings and more especially of the tendo-Achilles can be corrected by this means. When the cast is removed passive motion is used and action gradually taught. An attempt is made to so strengthen antagonists, during the period of partial incapacity of the tenotomized agonist, that the tendency for the contracture will be overcome. Working on this hypothesis Allison and Schwab[72] recommended the muscle group isolation by means of alcohol injection into peripheral nerves supplying the anteyonist. Foerster[73] recommended cutting of the posterior roots subdurally for the same purpose. According to him the posterior roots conveying sensory impulses from the periphery and allowing them to act on the cells of the anterior horns were an essential arm in the structure whose function it is to keep up tonus. By cutting the posterior roots of $C_{4, 5, 7, 8}$ and D_1, for the arm and $L_{2, 3, 5}$ and S_2 for the leg, he got a flaccidity but not a total inhibition of all sensory impulses from the limb. Kuttner[74] (Tr. International Congress of Medicine viii*a* (1913)) stated that in only five of 100 cases did he fail to improve the condition in the lower extremity by means of Foerster's rhizotomy.

Schüller[75] recommended section of the posterior columns of the cord. Sufficient data are not yet at hand to give this procedure its proper value.

The treatment of these cases, of course, varies for each case. Much can be done for the helpless idiots if sufficient patience and time are used. The most extensive reports of what may be done along this line are those of Bourneville in the Archives de Médicine from 1900 on. He reports many improvements in cases of the hopeless and helpless idiot showing that they may be taught to be tidy, to feed and dress themselves and even to talk in a fairly intelligent manner, at least enough to make their wants understood. With this improvement an improvement in temper is noted.

Special forms may require specific methods. For example: Hydrocephalus may be considerably helped with consequent improvement of the general picture. Frazier[47] divides hydrocephalus into four varieties. (1) Non-absorptus. (2) Obstructivus. (3) Occultus. (4) Hypersecretivus. The names are self explanatory. In the first variety he advocates drainage to the pleura. In the second, puncture of the corpus callosum, and in the fourth, the administration of thyroid extract.

In one case of moderate hydrocephalus of the hypersecretive type a most satisfactory improvement was seen immediately after the withdrawal of 5 c.c. of ventricular fluid through the anterior fontanelle. Irritability entirely disappeared so that the infant who had become impossible to handle immediately became perfectly normal in this respect. Muscular action and speech, which had shown no sign of improvement, soon came under voluntary control and rapid progress was made by the two year old boy. This progress was still satisfactory a year after the treatment, when the child was last heard of.

If the feeble mindedness is not due to syphilis or is not the consequence of a progressive sclerosis then much can be done towards adjusting the individual towards an environment in which he can make fair progress. It is a great mistake to seek to get normal progress out of an abnormal mind with the usual methods of education. It is a great mistake to put the subnormal child into competition with normal children. It is a theory, difficult of proof but, I believe, nevertheless true, that the unfairness and inequality of this competition is one of the causes of insanity, when it develops on the soil of feeblemindedness. Hence, the feebleminded child must be carefully guarded against too strenuous competition.

It is rare, indeed, that much can be done to aleviate the condition of the epileptic. Progress in the treatment of this condition must be made along lines somewhat on the colony basis. The epileptics must be put into an environment in which they can prove their usefulness, yet be cared for and protected at the critical periods. It is only under such conditions as these that any happiness is to be gotten out of life.

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CHAPTER CLXIX

SURGERY OF THE HEAD AND SPINE

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Introduction.—In the past decade the study of nervous diseases of children has received a great impetus through the rapid development of surgical methods of treating a considerable number of them. For this reason it is particularly desirable to consider such conditions primarily from a surgical standpoint and to group them together. There are numerous conditions which are not peculiar to children but which could not well be omitted in a comprehensive survey of the subject. Throughout the following chapters the principle emphasis will be laid on those conditions which occur in infants and adolescents and those conditions which, though they occur in adults, produce different manifestations in children.

Anatomical Features Peculiar to Children.—The scalp in children is relatively much thinner than in adults and is very loosely attached to the underlying structures. In this it differs markedly from the adult scalp. The periosteum also is loosely attached except at the suture lines where it is very adherent. These facts are of importance in explaining the spread of blood or pus through the scalp, and in accounting for the more rapid spread of infections over the scalp in children than in adults.

The skull is thinner than in an adult and contains less deposit of calcium. The outer and inner tables are more clearly marked; the diploic vessels are so prominent that they may be seen through the outer table at times when the scalp has been reflected.

The brain and meninges also show variations that are of great importance for diagnosis. The meningeal vessels which in adults often traverse the bone or lie in deep grooves in the bone, in children lie on the surface of the dura. Pacchionian granulations are entirely absent in infants and as the child reaches adolescence begin to appear. The longitudinal sinus has lateral prolongations which sometimes extend 1 to 2 cm. to either side of the median line. This peculiarity must be kept in mind if a ventricle puncture is undertaken as its neglect may lead to serious complications.

The characteristics of the child's brain have been described in the chapter on anatomy but a few points of surgical significance may be emphasized. The brain of the new born infant and during the first few weeks of life has a very different consistency and color than the brain of an older child. It is softer almost gelatinous, during the first few days and more translucent in appearance. This probably is due to the absence of myelinated fibers. The cortex, as seen at operation, looks much pinker than in older individuals but this may be due to greater hyperemia caused by the anesthetic. Due to the lack of development of the frontal lobes, the motor area lies relatively more anterior than in older individuals and the usual directions for outlining the central fissure on the skull can therefore not be followed.

In children under one year of age, there is a defect in the occipital bone in the region of the foramen magnum. The posterior portion of the foramen magnum is not yet formed and s represented by a membrane which is practically continuous with the atlanto-occipital membrane. This fact is of great value in operative procedures upon the fourth ventricle and posterior fossa, particularly in cases of obstructive hydrocephalus.

EXTERNAL COVERINGS

Congenital.—Nevi.—These may occur anywhere on the face or scalp. They vary in size and in the extent to which they involve the skin and subcutaneous tissues. There may merely be a discoloration of the skin without elevation above the surface or if the process is more marked the tumor may be elevated to such an extent as to cause considerable deformity. The color of these patches may vary from pale pink to blue or purple. When the process is elevated above the skin some treatment for cosmetic reasons may be indicated. Freezing the area with carbon dioxide sometimes removes small areas, at other times an electrolytic needle attached to the negative pole of a battery may be used to destroy these nevi. The needle with the current turned on is stuck into the mass parallel to the sufrace and kept in position for a minute or two. This may be inserted at several points. After a few days a scar has formed and the treatment is repeated.

These nevi, if they involve only the skin, are only of surgical significance for cosmetic reasons. Occasionally these superficial nevi are associated with a similar process intracranially. They may then cause focal epilepsy and require operation. The operations may be very difficult and the removal of the vessels, which at times are formidable both in size and number, may put the surgeon to a very severe test of his skill. The writer has described two such cases under the title of "Intracranial Telangiectasis." These nevi at times follow the distribution of the fifth cranial nerve. A large series of such cases has been collected by Marie and his pupils.

Traumatic.—The coverings of the head are frequently subject to trauma. Abrasions and incised wounds occur so frequently that practically no child escapes them. It is only when one has inspected many shaved heads that the frequency of these injuries is brought home to one. As a rule, the patient has no recollection of how many of these scars have been incurred.

They are *per se* of little surgical importance but in association with more deep seated trouble may deserve attention. A severe contusion of the scalp may lead to a subperiosteal hematoma which is difficult to distinguish from the irregularity found around a fracture. In infants the skin is so thin and loosely attached to the bone that infiltration of a large portion of the head readily occurs. There is no barrier between the attachment of the frontalis muscle anteriorly above the eyebrows and the superior curved line behind. And these two points may be the boundaries of such infiltration.

The old custom of differentiating scalp wounds into various types leads to unnecessary discussion. The points to be determined are: (1) Is the wound infected with ordinary pathogenic organisms and is there danger also of a tetanus infection? (2) How deeply are the tissues of the scalp involved, does the wound extend through the pericranium or only down to the galea? (3) To what extent has the blood supply of the scalp been jeopardized? (4) How long since the injury?

Incised wounds should be thoroughly cleaned, the hair shaved all around the wound edges and then a careful approximation made. A large poorly healed scalp wound may at some future time be a source of much annoyance. On several occasions such a poor scar has interfered with the blood supply of a cranial flap that was made for some intracranial procedure.

Avulsion of all or part of the scalp in children old enough to work in factories is a most distressing injury. Fortunately, in the past years greater precautions in factories have made these accidents less frequent. If portions of the scalp have been avulsed but the blood supply is intact they may be replaced with or without drainage. If the patient receives attention early before infection has made headway, the wound is to be treated as a contaminated wound, that is, *debridement* is to be practiced, the soiled areas are excised and the wound closed without drainage. If, on the other hand, an infection has started, it must be drained and in selected cases may be treated by the Carrel Dakin method. If the pericranium is not torn away with the skin a fresh granulating surface soon develops which can be skin grafted. When the blood supply of the skin has been destroyed, extensive areas of gangrene may develop and if in addition the pericranium has been stripped off, the problem of covering the defect may become a difficult one. In such cases the plan suggested by C. H. Mayo* is a good one. Numerous perforations are made through the outer table and from these buttons of granulation grow up and cover the denuded bone.. A Doyen perforator and burr are very satisfactory instruments for this procedure. It is well to remember that in children the skull is often so thin that the perforation may readily be carried through to the dura. Such a granulating area can then be skin grafted.

If there is any possibility of tetanus infection a prophylactic injection of serum should be given. It must be kept in mind that in many instances of scalp wounds the superficial wound is but an incident to much graver cranial and intracranial injuries. The treatment remains the same, however, though some modifications may be required.

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* Mayo, C. H., The preparation of dry bone areas for skin grafting, Ann. of Surg., lx (1914), p. 371.

Thus if the bone is fractured and the dura opened the debridement must include all these structures and under such circumstances the need for closing the defect in the scalp becomes imperative since drainage of intracranial lesions is never permitted.

Burns of the head present the same problems that they do when occurring elsewhere on the body. If the burn is of the first or second degree, cleanliness and a bland sterile ointment will suffice. If the burn destroys the entire skin, grafting becomes necessary after all sloughs have been removed and a clean surface has been obtained. At times burns destroy the pericranium so that necrosis of the bone may follow. The prevention of infection in such cases becomes of vital importance both because of the extensive bone necrosis that may occur and also because thrombosis of diploic veins may occur which may lead to a spreading thrombosis of the sinuses.

Inflammatory.—Abscess.—During the first year of life, the infant's scalp is constantly subjected to pressure and extreme watchfulness is necessary to prevent pressure sores with associated infection. The child's position has to be changed frequently and any redness of the scalp should receive prompt attention. Under-nourished children or those suffering from prolonged illness are particularly prone to get such bed sores. Such lesions may be superimosed upon a seborrheic eczema. From such an infection a troublesome furunculosis of the scalp may be started and as the tissues are so loosely attached at this age, the abscesses may become very large but be associated with little pain or discomfort. In pyemic cases following umbilical infection such abscesses may develop with great rapidity. They develop at points where the head presses on the pillow probably due to the slight anemia and consequent lowered resistance that develops at such points. In children with a large hydrocephalus such infections may cause much difficulty. Prophylaxis in these cases is the most important point in the treatment. Once the abscess has formed it must be freely incised. The time honored custom of using a ring to relieve pressure often aggravates the trouble. It is far better to have the head resting on a big pad of fine cotton which should be changed at least once a day. At times an infection of the scalp may take the form of a cellulitis.

Cellulitis.—As there are no fascial attachments to stop such an infection it may spread widely if it gets between the skin and the pericranium. Free multiple incisions are essential. In older children a not uncommon cause of infections of the scalp is pediculosis capitis. The removal of the pediculi is an essential preliminary step. From any of these infections the glands that drain the scalp may become involved. The posterior auricular group and glands of the posterior triangle of the neck are the ones most frequently affected and occasionally the submaxillary and parotid group become enlarged when the infection is in the anterior portion of the scalp.

Erysipelas.—This condition when it occurs on the scalp presents a somewhat different picture than elsewhere on the body. On account of the thick hair it may be difficult to make the diagnosis. The constitutional symptoms are present, fever, an initial chill and prostration with marked tenderness of the scalp. In fact, the pain is more marked in erysipelas of the scalp than in erysipelas elsewhere. This is due to the tough character of the skin of the scalp and its great thickness so that the tension of the spreading infection is greater.

The redness and sharp line of demarcation so characteristic of the disease may be entirely absent until the process has spread beyond the hair line. The differential diagnosis before this stage has been reached may be most difficult. The intense pain may prompt an over enthusiastic surgeon to consider surgical intervention; the knife and erysipelas are as incompatible as oil and water. There is no specific for the disease. Streptococcic serum and the countless ointments that are used are all of doubtful value. Cold compresses of sterile water are by far the best. The disease except in very weak, marantic children is rarely fatal though the clinical picture may be very alarming.

Tumors and Conditions Simulating Tumors.—Sebaceous Cyst (Wen).—This is the commonest tumor of the scalp. It may be single or multiple and at times reach an enormous size. In exceptional cases the scalp may be covered with them but this only occurs in older individuals. These cysts develop as a result of plugging of the duct of a sebaceous gland. They contain sebaceous material and broken down epithelial cells. They lie primarily in the dermis and extend down into the subcutaneous tissue but do not as a rule involve the galea. As they grow, the skin over them becomes stretched and thin and has a bluish appearance. Frequently in the center of the skin over the cyst may be seen the remnant of the plugged sebaceous duct. These cysts are not painful but may be annoying to patients in addition to their being very disfiguring. When they attain considerable size they readily rupture and become infected. When they are excised it is desirable to remove the entire wall of the cyst without rupturing it; leaving a small portion of the wall will lead to a recurrence.

Dermoids which have many of the clinical characteristics of sebaceous cysts usually are single and not multiple. They occur far more frequently in children whereas sebaceous cysts are found more often in adults. They develop at suture lines and at the site of embryonal clefts. They are more deep seated than sebaceous cysts and lie often beneath the galea, and at times cause erosion of the bone so that on *x*-ray examination thinning of the bone is observed and at times the bone may be entirely destroyed and the cyst lies in contact with the dura. Histologically they contain the structures of the skin. They may contain epithelium, hair or skin glands. They are difficult to differentiate from sebaceous cysts. They should always be removed as the thinning of the bone and exposure of the dura may give rise to intracranial symptoms, or if they rupture and become infected serious complications may arise.

Benign Tumors.—Lipomata and fibromata may occur. They are at best rare and particularly so in children. They have the same characteristics that these tumors have elsewhere in the body and require surgical treatment either for pain or cosmetic reasons.

Malignant tumors of the scalp may be either sarcomata or carcinomata. The latter more frequently arise in the non-hairy areas of the scalp or around the ear and spread up onto the scalp. As a rule they are epithelioma and are relatively benign. Radical local excision usually suffices.

Sarcoma on the other hand may be very malignant and rapidly involve the deeper structures. Under this heading must be included also tumors of the bone or meninges which secondarily involve the skin. The treatment must be determined by the extent of the involvement.

Occasionally metatastic malignant processes occur, of these the commonest, though also rare, are metatastic hypernephroma. They have a tendency to metastasize to bone and not infrequently select the skull as their particular point of metastasis.

Conditions Allied to Tumors.—There is a heterogeneous group of conditions that are included here which, though they are not true tumors, that is new growths, nevertheless give the symptoms of tumors. First among these are a variety of processes connected with the blood-vessels.

Angiomata.—These may be of three kinds, arterial, venous and capillary. They are all characterized by dilated vessels which are readily compressible and if of any size are bluish in color. They may cover a large portion of the scalp. At times these blood-vessel tumors are connected with the diploic vessels or the meningeal arteries. These dilated vessels may have extensive ramifications. In one such case seen by the author the vessels of the orbit and eye-ball were involved. Ophthalmoscopically it was possible to see dilated vessels lying free in the aqueous of the eye. When the process is as extensive as this, operative procedure is usually impossible; if, however, the mass can be ligated at its periphery, total extirpation may be possible. The surgeon should be prepared to deal with vessels passing through the bone and plug them with Horsley's bone wax. The various vascular conditions that have been described under this heading are *aneurysms*, usually of traumatic origin, *cirsoid aneurysms*, and *cavernous angioma*. The first of these, the aneurysm, develops in the course of an artery. The cirsoid aneurysm is a diffuse dilatation of an arterio-venous bed in which the arterial end of the bed is primarily affected as evidenced by the pulsation that characterizes this type of tumor. In the cavernous angioma there is no pulsation and in this condition the dilatation is primarily of the venous portion of the bed.

Pneumatocele.—This condition occurs rarely and is said never to have been observed in very young children. It is a collection of air under the periosteum and occurs in those portions of the skull where there are air containing cells—the mastoid and frontal sinuses. McArthur* has reviewed the literature up to 1905 when he reported his case and 32 others. In his table there were five cases in children under 15. Either from trauma or inflammation the cortex over air containing cells is destroyed and thus the air escapes under the periosteum. The treatment consists in laying the cavity wide open and allowing it to heal up by granulation so that the holes in the mastoid may be obliterated by granulation tissue.

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* McArthur, L. L., Pneumatocele of the cranium, Jour. A. M. A., Chicago, xliv, (1905), p. 1418.

Sinus Pericranii.—When first described by Stromeyer* in 1850 he had no understanding of its etiology or pathology. The term is really only of historic interest. In this condition we find a collection of fluid blood under the pericranium. It is really nothing other than a greatly dilated vein. Just as the superficial veins of the scalp and eyelids dilate generally when there is marked increased intracranial pressure, so in exceptional cases one vessel dilates inordinately. Cushing has only observed the condition in connection with brain tumors and in those cases it was an attempt on the part of nature to establish a collateral circulation since the intracranial vessels were compressed. The condition probably never occurs except where prolonged increased intracranial pressure has been present. Certainly when Stromeyer described the condition so little was known about brain tumors that it seems a justifiable assumption that all his cases were complicated by that condition.

Spurious Meningocele (Billroth's† Disease).—The name was given to the disease by Billroth. The term has been applied to collections of cerebrospinal fluid under the scalp but outside of the skull and the dura. The fluid pulsates synchronously with pulse and respiration just as in a true meningocele. The condition is differentiated from a true meningocele both by the history and its location. Bayerthal‡ gives a good review of the subject.

In every instance there seems to be some history of trauma. In order to bring about the condition there must be not only an injury to the bone but also to the dura so that the cerebrospinal fluid can escape. In very young children in whom the fontanelle is not closed I have seen the condition develop in the course of a puncture of the longitudinal sinus. In this case evidently the needle had traversed the opposite wall of the sinus and had entered the subarachnoid space. Compression of the scalp over the injury cleared up the condition. As a rule the condition clears up by itself and it would seem wiser to try such a conservative measure first. If the process has been present for a long time operation might be considered. The problem, however, is not as simple as it would appear. Evacuation of the fluid of course is of no avail. Closure of the defect in the dura may precipitate symptoms in a hitherto symptomless clinical picture. The continual excessive production of cerebrospinal fluid apparently affects the secretory powers of the choroid plexus so that a larger amount of fluid continues to be formed and thus intracranial pressure symptoms may readily be brought about if the defect in the dura is closed. It seems wiser therefore not to operate in this condition.

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* Stromeyer, Sinus pericranii, Deutsche Klinik (1850).

† Billroth, Th., Ein Fall von Meningocele Spuria cum fistula ventriculi cerebri, Arch. f. Klin. Chirurg., iii, p. 398.

‡ Bayerthal, J., Zur Kenntniss der meningocele spuria, Deutsche Med. Wochenschr., xxiv (1898), p. 37.

SKULL

Diseases of the Cranial Bones.—Rickets.—In this disease, due to defective metabolism, there is an insufficient deposit of calcium salts in the bone and a condition known as craniotabes develops. This is characterized by softened areas in the skull which are readily indented by the palpating finger. This is the only symptom of the disease which might bring it to the attention of the surgeon. Proper diet, if instituted in time, clears up the condition though as a result there may remain peculiar deformities of the skull. The head has a square appearance with unusually prominent frontal bossæ.

Microcephalus is a condition that no longer should be treated by the surgeon. The early closure of the sutures is not due to defective bony growth but is due to the maldevelopment of the cortex. The brain in these cases is primarily responsible for the early closure of the sutures. There is a microgyria always present with agenesis of the cortex. The early attempts of Lannelongue* to relieve the condition by making new sutures so that the brain might expand were misdirected efforts. Patients with this condition are usually idiotic and as far as we now know no surgical measures are indicated.

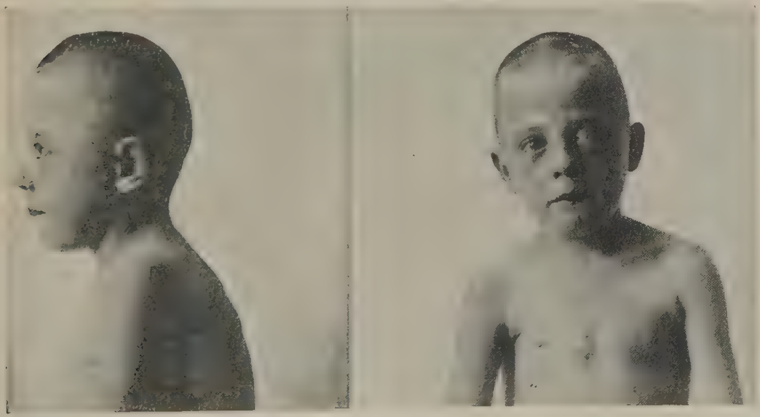

Fig. 1.—Oxycephaly. Typical prominence in region of the anterior fontanelle.

Oxycephaly (Turmschädel) (Figs. 1–3.)—This is a condition in which there is a prominence of the skull at the site of the anterior fontanelle from which the disease received its original name of turmschädel. The head in extreme cases has a dome-shaped appearance and seems compressed laterally.

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* Lannelongue, Bull. de l'acad. des sciences, June 30, 1890.

Etiology.—The cause of the condition is rather obscure and none of the theories are thoroughly satisfactory. It has seemed most probable to the writer that the fontanelles have closed prematurely in an individual whose brain was growing normally. This is quite different from what is found in the microcephalic where we are dealing with an abnormal brain which does not grow and consequently there is not enough intracranial pressure to keep the fontanelles open. In support of this theory, a study of tables of the growth of the child's brain indicates that the brain at the end of the first year has attained two-thirds of its full growth, while at the end of the fourth year it has completed four-fifths of its entire growth. The sutures of the skull are not solidly united until after the fifth year so that the growing brain readily pushes out the skull at its weakest points, the anterior fontanelle and the temporal fossæ.

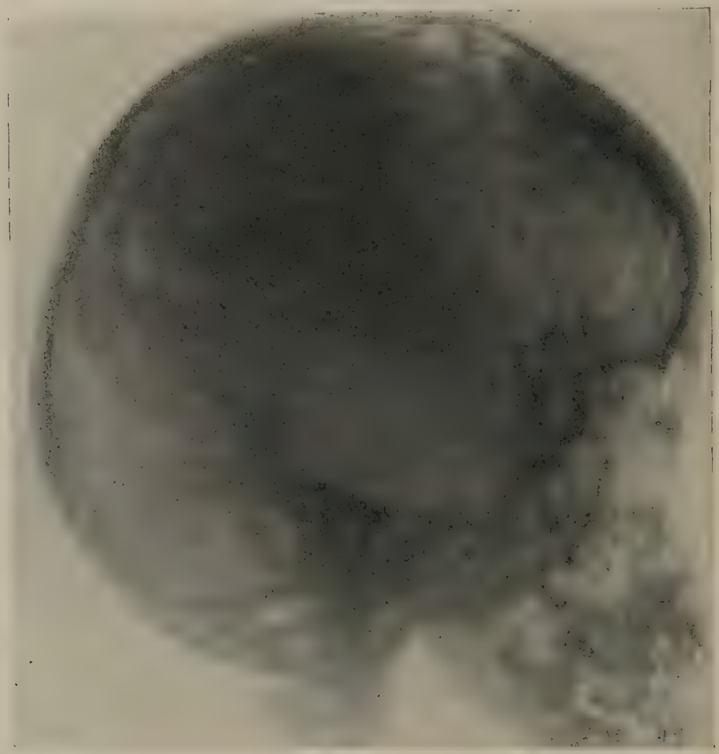

FIG. 2.—Oxycephaly. The extreme convolutional atrophy in a child is almost diagnostic of the condition.

Pathology.—The appearance of the brain in this condition is normal. The convolutions are well marked and show no variations from the normal. There is never any tendency to microgyria as in microcephalus. There are no recorded studies of the finer histology of such brains. The skull shows marked irregularities due to pressure atrophy. These irregularities are due to the pressure of the convolutions upon the bone and constitute what is so strikingly seen in the x-ray——convolutional atrophy. In addition to the general changes in the skull there is a marked prominence of the temporal fossæ which is apparent in an anteroposterior x-ray plate.

Diagnosis.—Headache usually is the outstanding symptom. In addition many patients have rather marked exophthalmos and complain of impaired vision. On further examination, a bilateral choked disc is frequently found. Examination of the skull reveals either an unusually high forehead or a prominence in the region of the anterior fontanelle which not infrequently only becomes evident when the patient's hair is closely cropped. The temporal fossæ, instead of being filled with muscle, are filled by a bony mass which is the laterally distended skull and in consequence the temporal muscles are in most cases poorly developed. The mentality of the patient is normal. There are rarely any of the other evidences of increased intracranial pressure. Vomiting, disturbances of respiration and drowsiness are characteristically absent. The cases that have choked disc, unless operated upon, ultimately develop optic atrophy and the resulting loss of vision often goes on to blindness. Every institution for the blind has a number of such cases among its inmates. There probably are far more cases than is generally supposed, that present the deform-

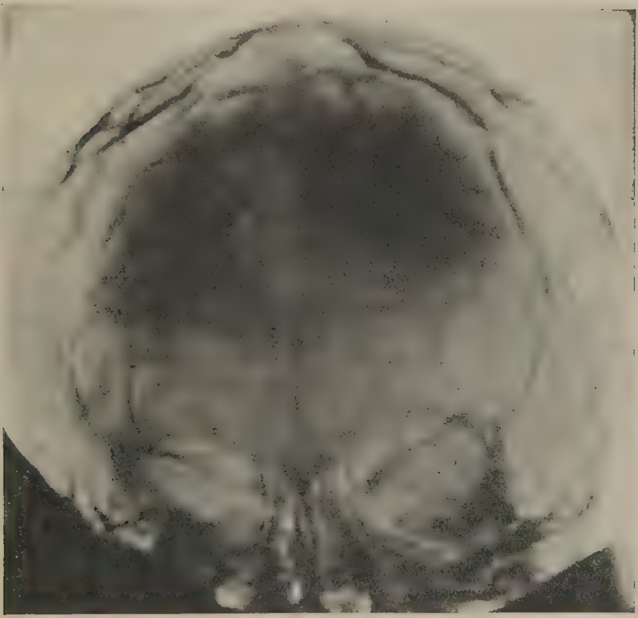

FIG. 3.—Oxycephaly. The extreme convolutional atrophy in a child is almost diagnostic of the condition.

ity of the skull but no symptoms. Dock* has recently made a study of a large number of negroes which indicates that many individuals with a hitherto unexplained exophthalmos have the characteristically prominent anterior fontanelle (when the thick hair has been shaved), and *x*-rayically show convolutional atrophy. He has offered no explanation of why this should occur more frequently among the colored race.

Treatment.—This has been well summarized by Sharp.† In those cases presenting pressure symptoms, either headache or choked disc, a unilateral or bilateral subtemporal decompression affords prompt relief. As the increased pressure is frequently very marked, opening of the dura, if it is at all tense, should be preceded by a ventricle puncture with the withdrawal of enough cerebrospinal fluid to reduce the pressure. From 10 to 50 c.c. may have to be removed. The marked prominence of the temporal fossa in these cases makes the operative procedure more troublesome than in a skull with normal configuration. This deformity of the temporal fossa also makes withdrawal of cerebrospinal fluid from the inferior cornu of the lateral ventricle difficult. The writer on several occasions has been unable to tap the ventricle at this point and has been obliged to withdraw fluid through a frontal lobe puncture.

Infectious Processes.—Osteomyelitis, when it occurs in the cranial bones, usually originates differently than in the long bones. In the latter, if not the result of a compound fracture, it always a is blood stream infection. When the disease affects the cranial bones it often begins as a periostitis or as an infection of a subperiosteal hemorrhage. In not a few cases an abscess of the soft parts involves the periosteal layer and by this route the infection gets into the bone. In young children the suture lines act as temporary barriers to the spreading of the disease. The infection may lead to necrosis of the outer table and in rare instances to necrosis of the entire bone. The infection may affect the meninges producing a fatal meningitis or it may affect the sinuses causing septic thrombosis which in turn may lead to septicemia and death.

Such a spread of an osteomyelitis may be looked for particularly if the disease begins in the mastoid and middle ear or in the sinuses of the nose. In the latter case it may with alarming rapidity perforate the thin ethmoid plate, produce an intraorbital abscess, and from there travel back and lead to a cavernous sinus thrombosis. Suppurating middle ear disease may also spread in young children with extraordinary rapidity. It is striking that sinus thrombosis occurs far more frequently in young children than in any other group of individuals. The greater vascularity of the bone and the much greater development of the diploic veins is probably the explanation for this.

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* Dock, G., Oxycephaly and Exophthalmos, Contributions to Medical and Biological Research, Osler Anniversary Volumes, Hoeber, i (1919), p. 433.

† Sharp, W., The cranial deformity of oxycephaly, Amer. Journ. of Med. Sciences, cli (June, 1916), p. 840.

The *treatment* consists in free exposure and drainage of the primary focus. If only the soft tissues are involved, drainage of the abscess suffices but if the bone is involved it must be removed to the outer limits of the infection. It may frequently be necessary to expose the dura. The appearance of the dura itself as well as the symptoms must be taken into consideration before the surgeon decides to open the dura in such an infected field. Unless he is certain that the infection has gone through the dura and that he is dealing with a brain abscess he should wait, clear up the superficial infection first completely and then explore the brain through a clean field.

Syphilis (Fig. 4) of the cranial bones both in hereditary and acquired syphilis is a common occurrence. The process begins as a periostitis and forms tender swellings which may be single or multiple. There may be bone proliferation and on examination a hard mass may be found that may be mistaken for an osteoma for in the later stage of the disease these swellings may no longer be tender. The masses are usually gummata and histologically show the same picture that gummata do elsewhere in the body. These masses may soften and break down though that is unusual for a syphilitic process unless they become infected. This, however, occurs not infrequently in children. The skin becomes adherent over the masses and a slight trauma

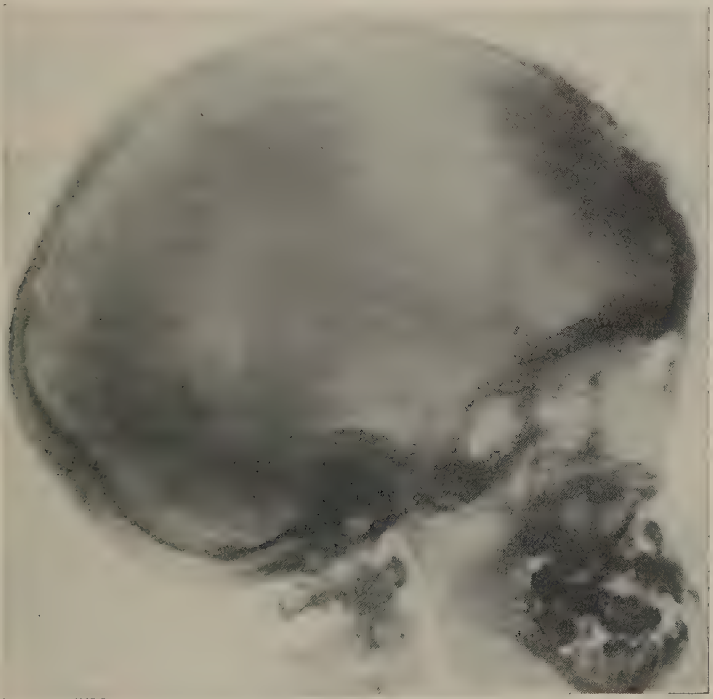

Fig. 4.—Syphilis of the skull.

or skin infection is enough to cause breaking down of the gummatous process. This may lead to very extensive necrosis of the skull with extensive sequestrum formation. Such a process cannot be cured by the administration of antisyphilitic remedies for the secondary infection has to be dealt with as well. Surgical measures are therefore essential. These necrotic areas should be cleaned up, the sequestra removed and often some bone that has not yet separated has to be removed as well. Care should be taken not to open new tissue spaces lest the infection spread further. Such wounds may be cleaned up more rapidly by Carrel Dakin solution, though care must be exercised that there is no lesion of the dura for Carrel Dakin solution in the subdural space is extremely irritating. These patients with syphilitic periostitis complain of intense pain in the early stages of the disease. At this time physical examination shows no abnormality of the skull but exquisite tenderness on even gentle palpation. Any antisyphilitic remedy, mercury or salvarsan, even iodids, relieves this acute pain in a few days so that one asks oneself the question whether the process has been in the bone at all or only in the dura.

Tuberculosis is prone to affect the bones in children and though it does not attack the cranial bones with the same frequency that it does the long bones, it nevertheless is seen occasionally. The disease has to be differentiated from syphilis and chronic osteomyelitis. Clinically it presents a picture so identical that it may only be possible to make the diagnosis by examining some tissue histologically or by animal inoculation.

The general appearance of the patient or the presence of other unmistakable foci elsewhere in the body may help in making a diagnosis. A point of importance may be the region in which the disease begins. Whereas syphilitic processes usually occur in the vault, tuberculous processes are more prone to originate about the nose or ear, that is orginate in mucous lined cavities. The presence of one or more fistulous tracts should always be enough to make one suspect a tuberculous process. The disease should be treated surgically in so far that any sequestra should be removed and the focus thoroughly cleaned out. This, however, constitutes merely the preliminary measures in the treatment; in addition the usual hygienic and dietary measures that are essential in treating any tuberculous case are to be instituted. Heliotherapy as advocated by Rollier* and others may be particularly useful in these cases.

Yaws, a rare disease in this country but is likely to be more common now that we have eastern possessions. The disease is produced by a spirochete resembling in many ways that of syphilis. The disease occurs in India, Japan, the Philippines and Hawaii. It has been studied particularly by Hutchinson,† Castellani‡ and Strong.§ The lesions are not very different from those of syphilis. Maul* has described cases of involvement of the cranial bones. I have seen a case in which the disease of the bones was very extensive and in which a cerebral abscess had developed. The disease, at least in this case, had not only eroded the bone but had also destroyed the dura in numerous places so that the brain herniated through the dura and lay directly under the skin. Salvarsan has the same effect on this disease that it has on syphilis. Bone lesions or a brain abscess as occurred in my case must be treated surgically.

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* Rollier, A., La cure de soleil, Paris et Lansaune (1914).

† Hutchinson, J., A Résumé of present knowledge as to framboesia syphilis, etc., Atlas Illust. Clin. Med. Surg., Lond. (1902), Fasc. 14, 3–27, pl. A to H.

‡ Castellani, Journ. Trop. Med. and Hyg., xviii (1915), p. 61.

§ Strong, Phil. Journ. Sci., Sec. B, v (1910), p. 433.

Fractures.—Just as elsewhere in the body, fractures of the skull may be divided into simple and compound, depending on whether the skin has been broken or not. Whereas in fractures of the long bones the first consideration is the injury to the bone itself, when we are dealing with fractures of the skull the fracture itself takes second place and the injury to the underlying structures becomes of prime importance. In the cases of compound fracture, infection is the great danger. The gravity of the situation is affected greatly by the condition of the dura. If the dura has been injured, the danger of the patient developing a meningitis or a brain abscess is far greater than if it is intact. This was strikingly seen during the war. The mortality among cases in which the dura was penetrated was far larger than in those cases in which the dura was intact. In children there are two sources for such infections, either following birth injuries or, in older children, following some trauma as a result of an accident. It is rather unusual to have a compound fracture following a birth injury. The author has seen one such case that was quite remarkable. The child, a year old presented the classical picture of hydrocephalus. The history was that the labor had been very difficult and during the extraction the head had been severely traumatized. There had been a scalp wound which had healed by granulation. Phenophthalein tests indicated an obstructive hydrocephalus. A median line cerebellar exploration (see hydrocephalus) was carried out. Some relief followed. The child finally died a year later. At autopsy an old fracture of the skull was found and extending in from this point a huge abscess occupying one hemisphere. The ventricle on the opposite side was dilated. All procedures of aspirating the ventricle and injecting phenophthalein had been carried out on one ventricle and consequently the pus had never been found. There was no suspicion that there was an abscess throughout the disease.

All fractures in which there is no break in the skin or mucous membrane are to be termed simple fractures. It must be recalled, however, that many cranial fractures may be compound, though appearing simple, because the fracture has torn some mucous membrane either in the nose, mouth or ear, and thus a portal of entry for bacteria is provided.

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* Maul, H. G., Bone and joint lesions of yaws with *x*-ray findings in 20 cases, Philippine J. Sc., Manila, xiii, Sec. B. (1918), pp. 63–77, 7 pl.

Mechanism of Production of Fractures.—Much has been written and much work has been done on the mechanism involved in the production of fractures. There are a host of workers, Kocher, Phelps,* Bergmann,† Cushing,‡ LeCount and many others. There are three general methods by which they are produced, by a bending, bursting or expansile force. It is often impossible in a given case to establish which one of these three forces has been at work, in fact it seems probable that at least two of the forces frequently take part in the production of a fracture, namely a bending and bursting force. Undoubtedly on account of the greater elasticity and softer character of the bone the factor of bending plays far more rôle in young children than in adolescents and adults. The lengthy discussions on this subject are primarily of historic and academic interest. Emphasis of this phase of the subject has rather tended to confuse the student and physician and has discouraged him in attempting to solve his problem. When the bone gives way at the site of impact we speak of a bending fracture while if the skull withstands the blow at the point of impact a more or less circular fracture or bursting fracture is produced. The factors which enter into the production of such a fracture are the severity of the impact, the tensile strength of the bone itself and to some extent the size of the surface through which the impact is delivered.

The bending force produces a depressed fracture at the point of impact but it is a very common experience to find radiating lines of fracture passing around the skull from the point of impact or from the edge of the depressed fracture. Whether these radiating lines of fracture are due to the bending force or whether there has been a bursting force applied as well has never been determined. The fact must be recognized that though the skull gives way at one point and a depressed fracture occurs with the resulting severe injury at that point such a case may also show the effects of a bursting fracture.

In those cases in which the skull does not give way at the point of impact a meridional fracture may occur; this may start at the point of impact or it may be through another diameter of the skull. In such cases the skull may give way at some thinner portion. Such fractures may pass through the base of the skull through any one of the three fossæ.

In dealing with fractures the cases may be classified into fractures of the *vault* and fractures of the *base,* though not uncommonly the two are associated. The fractures of the base are usually the result of a bursting force and, as the base is so well protected, a bending fracture practically never occurs there. The bending fracture occurs over the vault for there, though the bones are thicker, they are more exposed.

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* Phelps, Chas., Traumatic injuries of brain and its membranes, Annals of surgery (1906), p. 801.

† Bergmann, V., Lehre von den Kopfverletzungen, Deutsche Chirurgie Leipzig, (1880).

‡ Cushing, H., "Keen's Surgery," iii (1908).

The fractures due to an explosive force are produced by high velocity bullets which increase the pressure so tremendously and suddenly in the cranial cavity that the skull is shattered by the force from within. Countless instances of this sort have been recorded in the recent war. The cases are always fatal and therefore of little clinical interest.

Whereas what has thus far been said about fractures applies to children as well as adults in a discussion of the different types of fractures the age of the individual plays considerable rôle.

Linear fractures are breaks in the skull which are produced by the bursting effect of a blow. They extend around the circumference of the skull in an adult but in a child they are usually deflected by a suture line and then the imperfectly united suture line gives way and a diastasis of the bones occurs. In consequence, a linear fracture in a child may take on a very irregular course and produce an intracranial injury in a quite unexpected region. In children the dura is adherent to the suture line and when these fractures extend into the suture line they frequently tear the dura. Children therefore may more often be expected to have tears in the dura.

In depressed fractures the injury may involve only the outer table, only the inner table, or both tables. If both tables are fractured the inner table is more involved and a larger area of brain may be injured than one would expect as a result of the examination. Such fractures often are comminuted and it is in these that an injury of the meningeal vessels often occurs.

Perforating fractures are produced by some object of small size striking the skull with great force. Under this heading are included many war wounds, those produced by saber cuts, or bullets. A consideration of war wounds is not in place in a treatise on pediatrics. The gunshot wounds of civil life, however, must receive some consideration though they also are much rarer than in adults since suicide among children is so much less common than in adults. The accidental injuries are fortunately very rare.

Whereas the "temple" and the mastoid are the two common portals of entry in accidents in adults, the eye probably is by far the commonest entrance in children through the attempt to "look into the unloaded revolver." Consequently these bullets, if they enter the cranial cavity, lodge in the frontal lobes.

These wounds are attended by the same dangers that follow other compound fractures of the skull. The immediate dangers are due to hemorrhage, contusion and laceration with compression symptoms and sepsis. The late results from such a trauma may be mental deterioration, epilepsy, brain abscess or paralyses.

Stereoscopic *x*-ray examination should be made immediately to determine the position of the bullet. A probe should never be used as that only increases the danger of sepsis. If the dura has been penetrated, a *debridement* should be at once undertaken as outlined by Cushing.* This consists in removing the injured scalp and bone and cleaning out the bullet track with suction applied through a catheter. It is not necessary, in fact is often undesirable, to attempt to remove the bullet. This is particularly undesirable if the bullet has traversed the skull. The surgeon must consider each case as a separate problem and cannot treat these cases by rote. The experiences in the recent war, however, have made us change our views on the treatment of brain wounds. Prior to that time there was a strong feeling that they should be left alone, but today we believe that when a wound is still in the stage of contamination, and infection has not yet made any headway, it should be cleaned radically (debrided) and closed. The late complications of gunshot wounds must be treated regardless of the cause that has produced them, for these complications are not due to the bullet itself but the scar tissue which has developed in the track of the bullet.

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* Cushing, H., A study of a series of wounds involving the brain and its enveloping structures, Brit. Journ. of Surg., v (1917–1918), p. 558.

In trying to establish the diagnosis in any given case of fractured skull and outlining the proper treatment, the case must be considered from two points of view: first, the local condition of the fracture; second, the general and focal symptoms produced by the injury. Certain conditions in a fracture call for treatment irrespective of the symptoms arising from the nervous system. This applies particularly to depressed fractures. Whether the patient has symptoms or not, a depressed fracture in itself is a definite operative indication. Such fractures are to be elevated and if the dura is torn any blood clots or destroyed brain tissue is to be washed out very gently with warm normal salt solution and any bleeding vessels are ligated or preferably clipped with a Cushing silver clip.

Every case of compound fracture of the skull, even without symptoms, is to be cleaned out (debrided). If the dura is torn and the brain lacerated the debridement may also have to be carried into the brain substance. There may be an occasional case where the surgeon deems it safe not to do a debridement or unwise to undertake such a formidable procedure. The outcome of a policy of waiting, even "watchful waiting," is, however, more likely to lead to disaster. The surgeon feels a sense of security because the patient presents no symptoms. The harmless insignificant skin wound heals up, but in a few weeks or months or years the patient may succumb to a brain abscess.

There remains a large group of cases with fractured skulls which has general or focal symptoms due to intracranial involvement.

This intracranial involvement may be due to pressure from hemorrhage. If there is a steady rise in blood pressure it is evidence that a progressive hemorrhage is going on, and it is in order to recognize such a condition that every suspected case of fracture should have his blood pressure taken at frequent intervals. If the evidence points to a hemorrhage it behooves the surgeon to act quickly. A neurological examination may then reveal the side on which the hemorrhage is taking place. A paralysis of hand, face or leg, or even only paresis of one of these, may give the surgeon the clue he needs to determine the site of his exploration. At times a unilateral Babinski reflex may suffice. It is all important to remember that these neurological symptoms must guide the surgeon and not the location of the fracture. For example: A patient may receive a blow on the right side of the head and have a rightsided paralysis; the lesion causing these symptoms is undoubtedly over the left cerebrum and even if the facture on the right side is a depressed one the lesion causing the paralysis must be over or in the left cerebral hemisphere. Instead of the symptoms being paralytic in character they may be irritative, that is, the patient may have unilateral or focal convulsions (so called Jacksonian fits). These convulsions have the same localizing value and should indicate to the surgeon where the lesion is.

Certain other cases of fracture present this picture. They are unconscious with a blood pressure that does not vary and is normal or a little above normal. The patient may have stertorous breathing, there may be some abrasions of the scalp, but no fracture of the vault to be made out. These patients have a fracture of the base with contusion and laceration of the brain and edema of the brain rapidly supervenes. They may have a difference in their reflexes which would be of value in determining where the procedure to be described below is to be carried out. About the treatment of this group of cases there has been the greatest amount of discussion and the greatest difference of opinion. These patients all have greatly increased intracranial pressure due to the edema which develops as a result of the contusion and laceration. A subtemporal decompression operation may provide enough relief to tide the patient over this critical period. Some of these patients will inevitably succumb but the severe laceration of the brain is responsible for their death and not the operation. The author is convinced that certain of these patients who otherwise would die will be saved by such a procedure.

There are many cases that have undoubted fractures of the base or even linear fractures of the vault on whom no immediate operation should be performed and on whom a waiting policy is desirable. The cases that are semi-conscious and have no focal signs present no indications for operation and should be left alone. Prolonged rest in bed with moderate catharsis is all the treatment they need.

The cases with basal fractures have blood in the spinal canal. Cushing suggested some years ago that lumbar puncture would help to establish the diagnosis. This is quite correct but the procedure is attended by grave danger. I have seen a child die almost instantaneously after the removal of a few drops of fluid. The increased intracranial pressure when the cerebrospinal fluid is removed forces the cerebellum through the foramen magnum and causes a medullary collapse. Most of these accidents are not reported. Schwab and the author* some years ago investigated this question and found numerous instances of such an accident. Schönbeck,† has reported a considerable number of these cases from the German literature.

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* Sachs, E. and Schwab, S. I., Journ. A. M. A., lxvii (1916), p. 1651.

† Schönbeck, O., Die Gefahren der Lumbal punktion, Arch. f. Klin. Chirurg., Berlin, cvii (1916), p. 309.

The diagnosis of a fractured skull can practically always be made without the aid of lumbar puncture. The great improvement in roentgenology in the past 20 years since Cushing made his suggestion makes such a procedure unnecessary, furthermore, I have never seen a case whose treatment or prognosis was in any way affected by the presence or absence of blood in the spinal canal.

Very recently a new procedure has been described which may prove of great value in controlling the edema of cerebral contusion and laceration. Weed and McKibben* found that intravenous injections of saturated salt solution (35 per cent) caused shrinkage of the brain bulk. The author has tried this on several occasions and has found it so satisfactory that he believes it may replace many operative procedures for reduction of increased intracranial pressure occurring with fractured skull.

Late Complications of Fractures.—There are a series of conditions that may all be grouped under this general heading because they have a common etiological factor though they are very dissimilar.

Headaches.—One of the most frequent and most distressing symptoms observed after a fractured skull is headache. This may be so persistent and intense that it completely incapacitates the patient. It is usually not localized but is described as diffuse and not associated with anything the patient may do. Physical examination of these patients is entirely negative. There are no signs of increased pressure, the eye grounds are normal, the pulse is not slow, and the entire nervous system seems normal. As a rule the headache gradually disappears spontaneously in the course of a few months. This may be hastened or aided by rest and catharsis. Occasionally these palliative measures are of no avail and then something more drastic is called for. A number of these cases have been helped by subtemporal decompression. I have operated upon a number of cases of this sort and in every instance the dura has been tenser than normal, the pia arachnoid contained more fluid than normal and sometimes was slightly turbid. The release of this fluid combined with a decompression affords relief in some of these trying cases but the pathology is not at all clear; it seems probable, however, that the fluid in the pia arachnoid is not normal cerebrospinal fluid but is serum which is the remnant of an old blood clot not completely absorbed. The slight increased pressure produced by this accumulation of fluid seems to be responsible for the headache.

Dizziness is an even more distressing complication because there seems no satisfactory means of relieving it. This symptom is brought on most frequently with sudden change of position. Unquestionably it causes these patients much distress and incapacitates many of them. They are among the most distressing cases the neurological surgeon sees for they are outwardly well but totally unable to carry on even ordinary pursuits. Because their physical examination is quite normal they often receive scant sympathy or attention from the physician and this in the case of children leads them to become morose and not to mingle with other children of their own age.

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* Weed and McKibben, Experimental alteration of brain bulk, Amer. Journ. of Physiol., xlviii (1919), p. 531.

Epilepsy.—A certain percentage of cases, years after their injury, develop epileptic convulsions; though occasionally these are focal in character, as a rule they are general. A minute study of such cases reveals nothing abnormal, occasionally there seems to be a disturbance of pituitary function and if in addition, as often happens, the *x*-ray plate shows a deformity of the sella turcica, particularly the dorsum sellæ, the convulsions are laid at the door of the pituitary gland. Such a conclusion seems as yet unwarranted. Glandular feeding in such cases affords no relief though this is not an entirely satisfactory test since the methods of glandular feeding have not been stabilized. In the first place the extracts vary greatly in their physiological effect, and secondly, we have little conception of the proper dosage. Some experiments of Goetsch indicate that our dosage is entirely too small. The pituitary gland seems doomed to receive the same abuse and be misused as was the ovary by the passing generation of gynecologists.

The focal epilepsies all should still be given the benefit of surgical operation even though many of them will not be benefited by operation. Horsley, Kocher, Gerster, B. Sachs* and others have written much on this subject. At operation some of these cases show local scars, others show a diffuse degenerative process of the cortex. The cortex in such cases looks yellow instead of pink as does a normal cortex *in vivo*, there are areas where the cortex is destroyed and in its place there are cystic cavities varying in size from a pin head to cavities as large as a cherry or even larger. These cavities do not have a definite cyst wall. They have been variously described as arachnoiditis, and by others as subarachnoid cysts. The latter term carries with it the wrong connotation as these are not true cysts.

Victor Horsley's† doctrine still holds true that no focal epilepsy has been given every opportunity to get well until it has been explored. With the much smaller operative risk attending such explorations the procedure should continue to be recommended. The error that has often been made is that the procedure has not been carried far enough. Where there is a definite focal character to the epilepsy the cortical focus originating the fit should be excised, preferably by a subpial resection in order to obviate further scar formation.

Spastic Paralyses.—As a result of the cortical injury some fracture cases may have a residual paralysis. This may take the form of a monoplegia, diplegia or quadreplegia. At times such patients may have focal epileptic convulsions which begin in a paralyzed extremity or the convulsions may be general. Where there are convulsions the destruction of the cortex has not been complete and there is an irritative lesion which produces these convulsions. With these symptoms there often are associated varying degrees of mental deterioration.

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* Sachs, B. and Gerster, A. G., The surgical treatment of focal epilepsy, Amer. Journ. of Med. Sc. (October, 1896).

† Horsley, V., The surgical *vs.* the expectant treatment of intracranial tumors, Address before German Society of Neurologists, Berlin (October 6–8, 1910).

The *mental deterioration* may be of any grade of severity. In some cases the child is merely a little backward in school, in others it is a low-grade imbecile. These cases have been operated upon and at times evacuation of the collection of fluid under the pia arachnoid seems to help the mental condition but in most cases this is faulty observation on the part of the over anxious parent or the over enthusiastic surgeon.

These cases of diplegia or quadreplegia resemble clinically the so-called cases of Little's disease and often are so diagnosed. In fact, it frequently is quite impossible to tell them apart. There is a growing feeling among neurologists that they are one and the same thing (Freud,* Sachs†). Little,‡ it is true, considered prenatal injuries and asphyxia during labor the two important factors. In this he was correct but we know now that birth hemorrhages are much commoner than we formerly supposed and within the past few years Beneke§ has pointed out the much greater frequency of cerebellar birth hemorrhages which undoubtedly often produce the picture of quadreplegia.

One other complication that may be encountered is hydrocephalus. These always belong to the obstructive type of hydrocephalus and are largely if not always due to one of these tentorial tears described by Beneke. The blood gets into the fourth ventricle and closes up the foramina of Magendie and Luschka. The diagnosis is readily made, difficulty with nursing or breathing or both during the first few days of life followed by an apparent normal development. In a few weeks the head begins to enlarge suddenly. If there is no history of meningitis the diagnosis may almost be made on the history. Occasionally a cerebellar tumor or a congenital closure of the aqueduct of Sylvius, as described by Schlapp,‖ may produce the same picture. These cases lend themselves admirably to surgical treatment. A median line exposure of the fourth ventricle with opening of the roof and leaving the dura open permits the cerebrospinal fluid to escape into the subcutaneous tissue where it is readily absorbed.

Birth Injuries.—One cannot help but recall the words of the father of Tristram Shandie when one considers the great difficulties that beset the child upon its entrance into the world, "what havock and destruction must this make in the infinitely fine and tender structure of the cerebellum." The marvel is not that injuries to the head sometimes occur, but rather that they occur so infrequently. The compressibility of the bones of the skull and their pliability are factors that frequently prevent accidents.

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* Freud, S., "Die Infantile Cerebrallähmung." Nothnagel's Specielle Pathologie Vienna, ix (1897).

† Sachs, B., Contributions to the pathology of infantile cerebral palsies, N. Y. Med. Journ. (May, 1891).

‡ Little, On the influence of abnormal parturition on the mental condition of the child, Trans. of the Lond. Obstet. Soc., iii (1862).

§ Beneke, Ueber Tentoriumzerreissungen bei der Geburt, Muench. Med. Wochenschr., lvii, Hft. 2, p. 2125.

‖ Schlapp, M. G., Internal hydrocephalus with specimens showing occlusion of the aqueduct of Sylvius, Proc. N. Y. Path. Soc., n.s., xi (1911–1912), p. 64.

The commonest injury is the **caput Succedaneum.** The scalp is very loosely attached to the periosteum in an infant, consequently while the head is being molded, there is considerable pressure on the skull so that stasis of the blood and lymph circulation occurs and an edematous area develops on that portion of the protruding head on which no pressure is exerted. Depending upon the length of time the head is engaged as well as the amount of pressure that is exerted, the caput varies in size. It frequently is so large that the infant at birth looks greatly deformed but the condition is not serious and the fluid disappears in a very short time, usually not more than a day or two. The diagnostic signs are those of any edematous area, pitting on pressure but no fluctuation. The edematous area is diffuse and is not delineated by the edges of one of the cranial bones.

The condition never requires any surgical interference but is important because it must be differentiated from cephalhematoma.

Cephal Hematoma.—Some moderate trauma during labor, possibly through the application of forceps, leads to a separation of the periosteum from the bone and to a rupture of some of the vessels at this point with a consequent extravasation of blood. Whereas the caput succedaneum is at its maximum size immediately after the birth of the child, the cephal hematoma may not be present at all at birth and appears only after several hours or even days. It usually is unilateral and is delineated by the edges of the parietal bone; occasionally it is bilateral and sometimes over the occipital bone.

At times these hematoma increase in size for a few days and then gradually become absorbed in the course of about two weeks. If they are not absorbed at the end of that time, the clot may become organized into a mass of connective tissue or even have bone formed in it which may make a permanent deformity. At other times the clot may liquefy, break down and become infected. In order to avoid this latter danger, it is better to make a stab incision at the most dependent point of the tumefaction, express the clot and then apply a tight bandage for a few days. It is important to bevel the incision rather than make it straight as it will then close of itself and act as a valve. It is well to keep in mind that such a simple hematoma may have associated with it a fracture, and in order to avoid the very grave dangers from an infection in such a case, strictest aseptic precautions must always be observed. Obstetricians would do well to have this operation done more frequently.

Fracture.—Though the bones of the newborn are very thin and pliable, they are at times fractured during labor. The factors that may lead to this are prolonged labor with a contracted pelvis or a difficult forceps delivery. If the head in the process of molding impinges on some irregularity of the bony pelvis, the skull may become fractured, or if there is as yet very little deposit of salts in the bone so that it is very pliable, it may become indented. I have once seen a fracture produced by pressure from the small knob by which the traction arms are attached to a forceps. The extraction had been a very difficult one and in dragging the head through the pelvis too much pressure was put on the head. These fractures may at times be responsible for that most serious of all birth injuries, intracranial hemorrhage.

Intracranial Hemorrhage.—As a rule, however, this is brought about in a different way. Whereas fracture may be caused by poor obstetrics, an intracranial hemorrhage practically never is due to any faulty obstetrical procedure. These birth hemorrhages occur as frequently after an easy labor as after a difficult one, after a breech as well as a head presentation. As the head passes through the parturient canal the parietal bones overlap. This occurs in almost every case except those in which the head is extremely small or the pelvis very large.

If the head suddenly is born the springing apart of these overlapping bones leads to rupture of some of the veins entering the longitudinal sinus. These vessels may also be torn if the head has been markedly compressed so that the bones were overlapped to an extreme degree either by passing through a small pelvis or by the pressure of forceps; marked asphyxia may play a rôle but it has always been my feeling that since the ruptured vessel is one of the veins entering the longitudinal sinus, the principal cause of this grave accident is the springing apart of the bones when the head is suddenly released, combined with the fact that the veins passing from the cortex to the sinus are for a short distance quite unsupported and therefore much more readily injured.

The hemorrhage may consist of a thin film of blood which infiltrates the pia arachnoid or it may be a large clot, which covers one or both hemispheres. On account of the great distensibility of the child's head, an extraordinary amount of hemorrhage may occur without bringing about a fatal outcome or any symptoms comparable to those observed in an intracranial hemorrhage in an older individual with closed sutures.

The newborn child has so much of its nervous system still undeveloped—most of the cortex does not functionate nor do the pyramidal tracts which are still nonmedullated—and besides does not have its brain encased in a closed box, that practically all the symptoms due to increased intracranial pressure that are found in older children or adults are totally lacking. The most striking sign, if the hemorrhage is over the cerebral cortex, is the bulging bluish tense anterior fontanelle. Inspection of a large number of newborn infants shows that the anterior fontanelle normally never bulges except when the infant cries.

Stimulation of the infant produces no crying at all or at best a weak rather subdued response.

The sucking reflex which is always normally present at birth may be entirely absent or so weak that after a few ineffectual movements it ceases. At times the respirations are irregular and sometimes are typically Cheyne-Stokes or Biot in character. There may be cyanosis at times or a dilatation of the vessels of the scalp and eyelids due to increased intracranial pressure. I have seen a number of infants with unilateral convulsions and was formerly inclined to attach some importance to this symptom but believe now that unless there are other evidences one is not justified in making the diagnosis on this alone. The most valuable sign is the tense fontanelle. Whereas hemorrhage over the cerebral cortex may be difficult to recognize, a hemorrhage in the posterior fossa offers even greater difficulties. In these cases the anterior fontanelle does not bulge at all as the rigid tentorium cerebelli confines all the pressure to the posterior fossa. The most striking symptom, frequently the only one, is respiratory disturbance.

The history of the labor may be a further aid in the diagnosis though physicians should remember not to be misled by the history of an easy uncomplicated labor. But the determination of the side on which the lesion is will always remain a most difficult one so that at times a bilateral operation may be necessary.

Were all children with birth hemorrhages, if untreated, to die, one might only suggest what under the most favorable circumstances is a somewhat formidable operation but a large number of these infants survive and form one of the most distressing groups of cases, and therefore the operative treatment should be insisted upon.

The later effects of this condition are numerous. All grades of mental deficiency may result, varying from the low-grade imbecile to the backward child. Cerebral birth palsies and what has often been erroneously called Little's Disease may be late results of these birth hemorrhages.

Epilepsies, both general and focal, may occur, and occasionally an obstructive hydrocephalus due to closure of the Foramen of Magendie by adhesions, the result of a hemorrhage. I have seen one such case and believe that not a few obstructive hydrocephalus cases may have this origin. The case in point was seen two days after birth with the classical picture of birth hemorrhage. Operation was refused and a year later the child appeared with an obstructive hydrocephalus which was relieved for about two years by making an artificial Foramen of Magendie. Recently Beneke has pointed out that in the past many birth hemorrhages have been overlooked. In examining a large number of stillborn fetuses he found that many of them had a posterior fossa hemorrhage due to a tear of the tentorium of the cerebellum. The reason these had heretofore been missed, he believes is due to the method of opening the skull and he has therefore devised a new technique for this purpose. Doctor Burrows at my request examined a considerable number of fetuses and confirmed Beneke's findings. Since my attention has been drawn to this I have seen a number of children in the pediatric clinic whose symptoms could readily be explained by an old posterior fossa lesion. I refer to the cases of birth palsies in whom incoördinate movements are the most prominent feature and spasticity is not so marked. Whether it will be possible to recognize these cases early enough to evacuate the clot before damage is done, only the future can tell.

The treatment of intracranial birth hemorrhage was first suggested and carried out by Cushing.* He pointed out in the first place that a cranial operation is well tolerated if the body temperature is maintained. He advocated turning down a bone flap and dural flap and washing out the clot with warm saline solution. For this operation he devised an admirable bone forceps which is very serviceable. Of course if this instrument is not at hand a pair of heavy scissors will do. He has further emphasized the importance of a careful closure of the dura and scalp. The infantile brain has a tendency to become more edematous than the adult brain and this may cause difficulty in closing the dura. This can be readily obviated by a withdrawal of cerebrospinal fluid, preferably by lumbar puncture, for the cortex is so gelatinous in character in infants that even a needle puncture may cause considerable disturbance.

The anesthetic should be carefully administered, a few drops of ether sufficing for the procedure.

In addition to this, I consider it very important to give all such patients subcutaneous salt before operation. An infant a day old may readily absorb several ounces. This is a most essential step of every cranial operation carried out on a child and one that influences the success of a procedure greatly.

More recently Simmons† has suggested a different operation which has many advantages. Instead of turning down a flap, he makes a small incision, not more than 1 or 2 in. long at the outer margin of the anterior fontanelle. The dura is exposed and incised. If a blood clot is found it will be extruded by the increased intracranial pressure. If the lesion is not found a similar procedure can be undertaken on the other side.

This is so much simpler than turning down a flap that it has much to commend it. Furthermore, in doubtful cases it could be safely employed as an exploratory porcedure.

It can be carried out without a general anesthetic by merely infiltrating the skin with salt solution.

In those cases in which the skull has been indented, a policy tempered with conservatism should be adopted. There is no doubt however, that at times the neglect of such an indentation may give rise to disturbances in later life. It may, through pressure, start an epilepsy or be responsible for a picture such as is seen after hemorrhage. A certain number of these depressed areas correct themselves, that is, the bone is pressed out into shape by the growing brain. If it does not correct itself in the course of a few days, a staphylorrhaphy dissector may be introduced through a small trephine opening and the depression raised. If the depression is over a silent area operation is not called for unless the depression is disfiguring, in which case the operation may be demanded for cosmetic reasons.

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* Cushing, H., Concerning surgical intervention for the intracranial hemorrhages of the newborn, Amer. Jour. of Med. Sc., cxxx, p. 563.

† Simmons, C. C., Two cases of intracranial cerebral hemorrhage, Bost. Med. and Surg. Journal, clxvi, p. 43.

BRAIN AND MENINGES

Anatomy and Physiology.—The coverings of the brain consist of the dura, the arachnoid and the pia mater and the lining of the ventricles—the ependyma.

The dura is a very thick tough membrane which completely envelops the brain and is attached to the edge of the foramen magnum and at the various foramina of exit of the cranial nerves. It is adherent to the inner surface of the skull and in young children is very adherent at the suture lines. It has as a prolongation the falx cerebri which lies between the two hemispheres and the tentorium of the cerebellum so that the cranial cavity is divided into three compartments. It is lined with endothelial cells and has the power, in the young at least, of producing bone; occasionally it assumes this function in adults and may be a source of confusion in the interpretation of x-ray plates. The dura is so tough and little distensible that it interferes as much with any expansion of the brain as does the bone itself, in fact, the dura does not give way to pressure as readily as does the bone. This is an important factor to keep in mind when intracranial pressure is to be relieved. *A satisfactory decompression operation must always include a large opening in the dura.*

It contains arteries which are branches of the middle meningeal artery. As this vessel passes up from the foramen spinosum on to the dura it lies in a deep groove and this groove is often bridged over so that when an osteoplastic flap is turned down it may readily be injured. It is at this point also that the vessel is injured in fracture or at the point of its passage from the foramen spinosum on to the dura. At this point the vessel has little support and a sudden jar may snap it off.

The veins in the dura are large sinuses. The longitudinal sinus lies in the median line, at times slightly to one side. It is narrow anteriorly and broadens as it passes back to enter the torcula Herophilii. In its middle portion in the region of the anterior fontanelle it has great lateral prolongations which are much more marked in children than in adults. These lateral extensions (parasinoidal sinuses) extend out for 1 to 2 cm. and must be kept in mind when a ventricle puncture is done. As they lie between the two layers of the dura, the outer layer may be incised before one is aware of their presence. In young children Pacchionian granulations are entirely absent or but slighty developed. These lateral prolongations of the sinus are not as marked in older individuals. I have a definite impression that the more the Pacchionian granulations are developed the smaller are the lateral extensions of the sinus.

The principal veins of the cortex enter into the longitudinal sinus. It is at these points that the vessels give way during labor and lead to a birth hemorrhage.

The Pacchionian granulations extend up into the sinus and have to do with the passage of waste products into the circulation as has been shown by Weed,* Wegefarth and others. Nevertheless the exact function of these granulations has not been worked out nor what their relation is to the cerebrospinal circulation. These granulations are often imbedded in deep pockets in the bone and are readily injured when the bone is removed. Any surgical procedure extending close up to or across the sinus is always attended by more or less troublesome hemorrhage. After the longitudinal sinus reaches the torcular it branches into three sinuses, two large lateral ones and a smaller occipital one which passes down the median line to the foramen magnum where it joins a circular venous sinus which is part of the spinal circulation. The lateral sinuses pass outward to the mastoid region where they dip down to form the sigmoid sinus which ends in the jugular bulb which in turn is the beginning of the internal jugular vein. The petrosal sinuses extending from the lateral sinus only require mention here; they may be encountered in operations on the Gasserian ganglion but if carefully handled do not cause any trouble.

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* Weed, L. H. and Wegefarth, P., Studies on cerebrospinal fluid, Journ. of Med. Research, Boston, n.s., xxvi (1914–1915), pp. 21–176.

Of very great significance, however, is the cavernous sinus which lies around the sella turcica and into which the ophthalmic and orbital veins empty. Any obstruction to this sinus causes stasis and an extraordinary degree of exophthalmos. This is particularly seen in cavernous sinus thrombosis. To what extent interference with the circulation of the sinus has to do with choked disc is a point of great interest. Not only is this important in reference to intracranial conditions but in those obscure conditions that come in numbers to the rhinologists complaining of headache and with changes in the eye grounds. Sluder* has devoted much time and thought to these conditions and believes at least in some of the cases the disease (usually inflammation) passes from the nasal sinuses through the bone to the optic nerves and no doubt at times to the cavernous sinus.

The *arachnoid* is a thin membrane encasing the brain loosely and underneath it is the *pia mater* which covers the cortex. The larger vessels of the brain lie in the space between the arachnoid and pia and the smaller ones lie in the pia entirely and furnish the blood supply to the cortex. These vessels run in the sulci and send lateral extensions over the cortex so that the center of a gyrus is practically bloodless and can be incised with impunity.

The ventricles of the brain and the choroid plexuses are covered by ependyma. Until very recently the view of Dixon and Halliburton† that the cerebrospinal fluid was secreted by the choroid plexus had become generally accepted. The work of Cushing‡ and his associates, and Frazier,§ all seem to support that idea. Recently, however, Becht‖ has thrown some doubt on all this work. His work is very careful but not sufficiently convincing to warrant throwing aside the whole conception. There is no doubt that the theory that cerebrospinal fluid is secreted by the choroid plexus, though still an hypothesis, has explained quite satisfactorily certain conditions that hitherto have been very obscure, such as the development of hydrocephalus after closing a spina bifida, the formation of a hydrocephalus after an obstruction in the region of the fourth ventricle, and the production of hydrocephalus in obstruction due to an inflammation.

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* Sluder, G., "Concerning some headaches and eye disorders of nasal origin." Mosby, St. Louis (1918).

† Dixon and Halliburton, Journ. of Physiol., xlvii (1913), p. 215; xlviii (1914), p. 128.

‡ Weed and Cushing, Amer. Journ. of Physiol., xxxvi (1915), p. 77.

§ Frazier and Peet, Amer. Journ. of Physiol., xxxv (1914), p. 268.

‖ Becht, F. C., Studies in cerebrospinal fluid, Amer. Journ. of Physiol., li (1920), p. 1.

Congenital Anomalies (Figs. 5, 6).—Practically all the anomalies are developmental in character and may be grouped under the general heading of *cephalocele.*

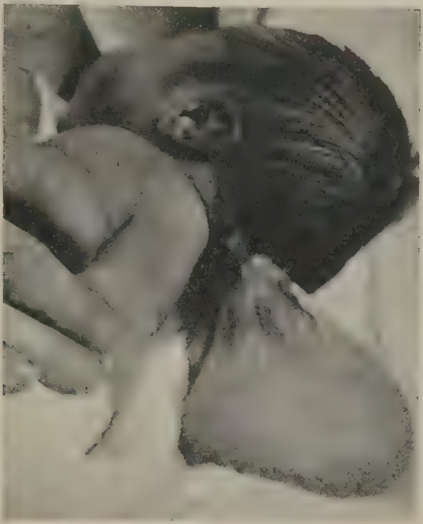

FIG. 5.—Occipital meningocele.

These are due to the failure of closure of the primary vesicles of the brain but this does not explain the fundamental underlying cause of the condition. In some cases there is a history of syphilis but this is by no means a constant finding. The defects in the spine (spina bifida) are identical in origin. I have seen one family in which three children in succession had spina bifida. Both parents were healthy and there was no evidence of syphilis. There had been one or two relatives who had also had children with spina bifida. Such cases strongly suggest a defective *anlage.* There are, however, other theories that have been suggested, none of which seem to be very probable. Berger's* idea that they are true neoplasms seems to have little foundation. The idea that amniotic adhesions are responsible, also does not seem very probable. It has always seemed to me that the underlying cause is a defective secretion (hypersecretion) of cerebrospinal fluid and that this excessive fluid keeps the neural groove from closing

* Berger, Des certaines Encephaloceles, Revue de Chirurgie, ix (1890), p. 162.

over at some point. This theory does not explain why in one case a sacral spina bifida develops, in another a dorsal, and in another patient a cephalocele. This idea that the seat of the trouble lies in the secretion of cerebrospinal fluid is borne out by those cases of spina bifida which develop a hydrocephalus following operation. Every surgeon has had these distressing sequelæ. I have had one case of occipital encephalocystocele that developed hydrocephalus after the defect had been closed by transplanted fascia. We must acknowledge that there are cases of this type (I group the spinal and cranial cases together) in which no hydrocephalus develops. Such cases might be brought forward against the secretion theory but this is not necessarily so for the disturbance in secretion may become adjusted in certain cases so that there is no more hypersecretion.

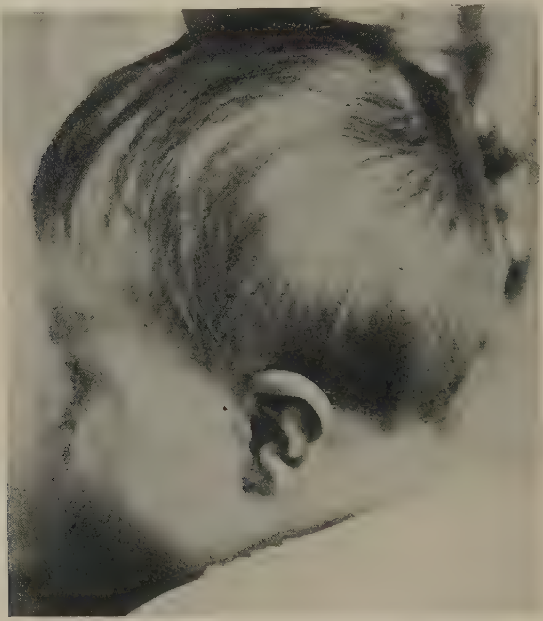

Fig. 6.—Occipital meningocele. Same patient 1 year after operation.

Varieties.—Cephaloceles occur in the median line and are spoken of as occipital or sincipital cephaloceles. They may occur above or below the tentorium. The ones below the tentorium, the inferior occipital ones often are connected with the foramen magnum so that it may be difficult to determine if the defect is spinal or cerebral.

The superior occipital encephaloceles may communicate with the posterior fontanelle, the anterior or sincipital encephaloceles protrude at the base of the nose and have had a variety of names applied to them according to the location of the defect—nasofrontal, naso-ethmoidal and naso-orbital.

There are three general types of cephaloceles:

Meningoceles in which there is only a herniation of the membranes without the prolapse of brain tissue.

Encephaloceles in which brain tissue prolapses.

Encephalocystocele in which there is a dilatation of the ventricle into the hernia and the coverings consist of the membranes and greatly thinned cortex. The diagnosis of this type may sometime only be made by microscopic section for the brain tissue may be so thinned out that nothing remains of it but the ependymal layer.

There are a large variety of more extensive developmental defects which may all be grouped under the heading of *Craniorrhachischisis*. They have been given various names depending upon the extent of the defect. The more common ones are Holo- and Mero-Acranic Anencephalie and Exencephalie. These are all of greater interest to the pathologist and embryologist than to the surgeon. Many of these children are nothing but monsters and either are born dead or die soon afterwards. Lysenkow* and V. Bergmann† have written excellent treatises on these cases. The true encephaloceles may vary greatly in size and at times are as large as the head itself. They may be pedunculated or sessile and at times are surmounted by a tumor, lipoma or fibroma.

The diagnosis is usually simple. The lesion is present at birth, is soft cystic in consistency. If the skin is very thin the tumor appears bluish. It pulsates and when the child cries becomes tense. If the tumor is compressed and the cerebrospinal fluid is forced into the cranial cavity, compression symptoms are readily produced. The crying child becomes quiet, sleepy and ultimately unconscious. The pulse becomes slow, and the pupils become dilated. V. Bergmann carried this dangerous and totally unnecessary experiment so far that he produced Cheyne-Stokes respiration.

At times it may be difficult to decide if the case is operable or not. V. Bergmann has stated the operative contraindications quite well:

1. If the head is flattened and small.
2. If the cephalocele is of the inferior occipital type and extends through the foramen magnum and includes the spine.
3. If hydrocephalus is already present.
4. If there are other deformities in the child which are likely soon to lead to its death.

The treatment if undertaken at all must be radical. Any attempt to obliterate the cavity by the injection of irritating substances is to be condemned. The sac must be excised and the defect closed either by a transplant from the infant itself or some other individual, preferably the mother.

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* Lysenkow, Hirnbrüche, Encephalocelen und ihre Heilung, Moskauer Dissertation (1896).

† Bergmann, V., "Chirurgische Behandlung von Gehirn krankheiten," Berlin (1899).

The defect may be closed by a piece of fascia or bone as advocated by Lysenkow and Albee. Trout used a homotransplant successfully in several cases of spina bifida. I have preferred using fascial transplants with the idea of not having such a firm closure.

Hydrocephalus.—An accumulation of cerebrospinal fluid in the ventricles or in the subarachnoid space. The condition may be acute or chronic. The type in which the ventricles are dilated (internal hydrocephalus) is far more common. External hydrocephalus occurs very infrequently and there is some question whether it is not a transformed internal hydrocephalus in which the fluid has escaped from the ventricles and accumulated in the subarachnoid space.

The old terms congenital and acquired hydrocephalus are of little value and it is far better to classify the cases according to the method of production of the hydrocephalus. The studies on the production of hydrocephalus by Thomas,* Blackfan and Dandy† and the work on the source of cerebrospinal fluid by Halliburton and Dixon,‡ Cushing, Weed and Wegefarth,§ Frazier‖ and Peet, and others, make it possible to classify them quite differently. Hydrocephalus may be considered under four headings:

1. Obstructive
2. Hypersecretive
3. Non absorptive
4. Mixed cases

The grouping into these types has been made possible by the work of Blackfan and Dandy. They showed that if phenolsulphonaphthalein were injected into the ventricle it would appear after a brief interval in the spinal fluid and that it would be excreted in the urine at a definite rate.

If there is an obstruction between the ventricles and spinal meninges the dye appears much more slowly in the spinal fluid. If there is a defect in absorption the dye appears very slowly in the urine. If there is no obstruction and no delay in absorption the dye appears promptly in the spinal canal and promptly in the urine.

The *obstructive cases* may have their obstruction at the aqueduct of Sylvius or in the roof of the fourth ventricle at the foramen of Magendie and foramina of Luschka (Fig. 7). This obstruction may be due to a tumor, to an inflammation as a result of a meningitis, the organization of a blood clot following a posterior fossa birth hemorrhage or a congenital anomaly (Fig. 8).

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* Thomas, W. S., Experimental hydrocephalus, Jour. Exper. M., Lancaster, Pa. xix (1914), p. 106.

† Dandy, W. E. and Blackfan, K. D., An experimental and clinical study of hydrocephalus, J. A. M. A., Chicago, lxi (1913), p. 2216.

‡ Dixon and Halliburton, Journ. of Physiol., xlvii (1913), p. 215; xlviii (1914), p. 128.

§ Weed, L. H. and Wegefarth, P., Studies on cerebrospinal fluid, Journ. of Med. Research, Boston, n.s., xxvi (1914–1915), pp. 21–176.

‖ Frazier, C. H., Types of hydrocephalus, their differentiation and treatment, Amer. Jour. Dis. Child., Chicago, xi (1916), p. 95.

In the *absorptive* cases there is some process which has affected the villi or pacchionian granulations through which the cerebrospinal fluid passes into the circulation and then is excreted in the urine.

In the *hypersecretion* cases the choroid plexus is presumably the seat of the disease and secretes more rapidly than normally so that though the pathways of absorption are intact they cannot dispose of the fluid rapidly enough.

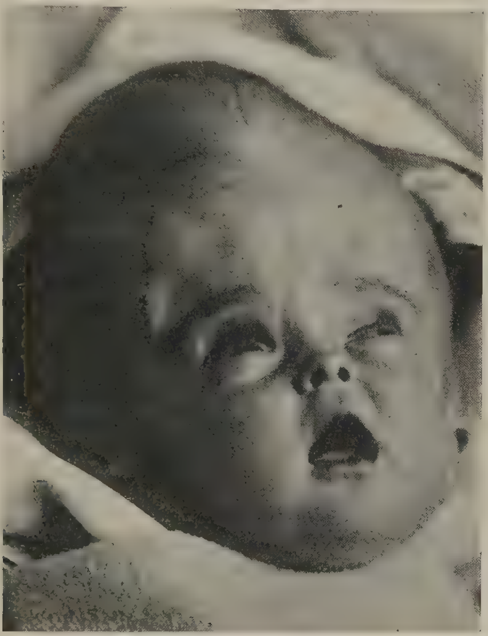

Fig. 7.—Obstructive hydrocephalus associated with a spina bifida which had become infected at birth and caused a basal meningitis.

Diagnosis.—In very pronounced cases the diagnosis is readily made; the huge head with prominent frontal bossæ, the eyes partially hidden by the protruding frontal bones, the sutures separated and the fontanelles wide open. These children are usually very fat and have accumulations of fat on their legs and bodies so that the parents exhibit them proudly because they are so well nourished. This symptom is evidence of pituitary insufficiency. The pituitary gland is always flattened out in these cases. These characteristic fascies may be seen in any type of hydrocephalus. The picture is more striking the earlier in life the condition begins especially if the sutures have not yet closed, but it may be very striking also in older children. I have recently seen a boy of eight years who developed an enormous hydrocephalus at the age of four following a pneumococcus meningitis. The presence of these hydrocephalic fascies must not be interpreted to mean that the condition has been present since birth and that the case is one of what was formerly called congenital hydrocephalus.

In addition to these symptoms, the patients may have a choked disc and paralytic symptoms or convulsive attacks. If the lesion is in the posterior fossa there may be cerebellar symptoms. The proper treatment of these cases can only be determined when we know what type of hydrocephalus we are dealing with. When that has been accomplished a rational procedure can be outlined.

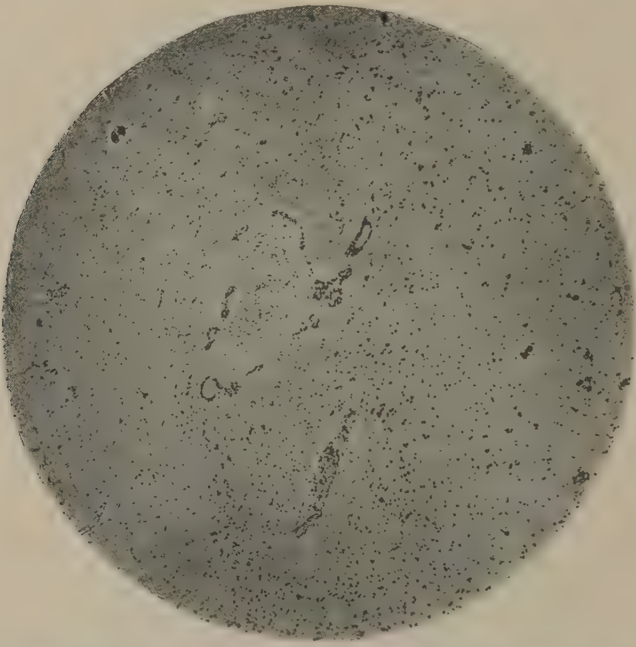

Fig. 8.—Congenital closure of the aqueduct of Sylvius which had caused an obstructive hydrocephalus.

If the case is one of obstructive hydrocephalus the region of the fourth ventricle and posterior fossa must be explored. This may be done by the usual cross-bow incision advocated by Cushing. I have for several years been carrying out such an exploration through a median line incision by retraction of the muscles without cutting them from their attachment to the occipital bone (Fig. 9). This does not give as extensive an exposure as the cross-bow incision but has proven very satisfactory as a preliminary measure. At any time the incision can be transformed into a cross-bow exposure. By this method the fourth ventrical is well exposed and an old basal meningitis may be dealt with or even a tumor of the middle lobe (vermis). By making a hole into the roof of the fourth ventricle and leaving the dura open, a pathway for the escape of cerebrospinal fluid into the subcutaneous tissue is afforded. The subcutaneous tissue is one of the best absorbing surfaces we have in the body.

If the hydrocephalus is caused by defective absorption the problem is to provide a channel for absorption of the cerebrospinal fluid. I have employed the same operative procedure in such cases.

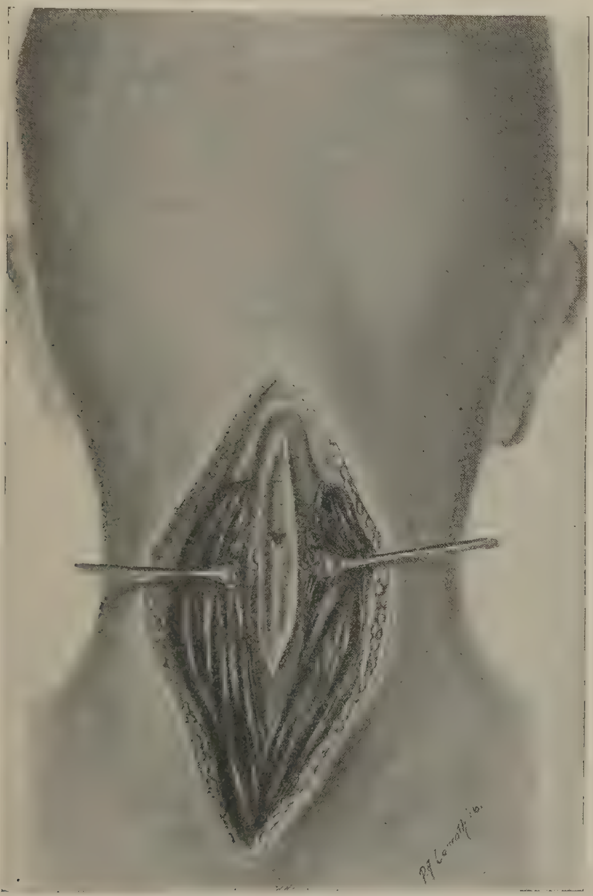

FIG. 9.—Median line exposure of the fourth ventricle for draining cerebrospinal fluid into the subcutaneous tissue.

If the condition is due to excessive secretion of cerebrospinal fluid the problem is very different. In such a case the attack must be made on the secreting gland. The rational treatment would seem to be to remove a part of the secreting gland just as is done in hyperthyroidism—part of the choroid plexus must be excised. This has been tried a few times by Dandy and several times by me. One case died a few days after operation and in the other one I was unable to expose the plexus as it was bound down by an ependymal inflammation. Some more recent cases have been relieved but in none did the hydrocephalus disappear. With certain types of choroid plexuses it may be a very difficult operation.

Many other methods have been suggested to treat hydrocephalus. All of them employ foreign bodies (tubes) to carry the fluid into cavities that would absorb the fluid. Frazier drains it into the pleura, Cushing by a combined laparotomy and laminectomy placed a tube through the body of the vertebra to drain into the retroperitoneal space; Kausch drained the fluid with a long rubber tube into the peritoneum. Cushing also drained from the subarachnoid space into the jugular by anastomosing a vessel; Horsley placed a gold tube from the ventricle to the subarachnoid space; and Payr connected the ventricle with the longitudinal sinus by means of a vein. All these methods have the objection which it seems to me my method has not, that a foreign body is used which soon becomes encysted and therefore soon ceases to drain. Some years ago puncture of the corpus callosum was suggested by Anton* to relieve this condition. The enthusiasm that first followed the announcement of this method has rapidly waned and most men have given it up. It does not attack the seat of the trouble and has only a palliative effect in a few cases. The treatment of hydrocephalus has not been solved as yet but it does seem as if we were making progress in a hitherto most disheartening field. The fact that a child has an internal hydrocephalus does not *per se* make the case hopeless for I have seen dilated ventricles return to normal size after proper drainage.

Cerebrospinal Leakage.—This is only a symptom, not a disease entity. Whenever there is an external discharge of cerebrospinal fluid some connection exists between the subarachnoid space and the exterior. This may be due to an injury; or when there is marked increased intracranial pressure as in hydrocephalus with or without tumor. The commonest site for such a leak is in the nose, *cerebrospinal rhinorrhea.* It may be an ineffective attempt on the part of nature to take care of increased intracranial pressure. Such cerebrospinal fistulæ are always grave cause for concern for they furnish a possible portal of entry for organisms and the production of a meningitis.

Cerebrospinal fluid may work its way through tissues and develop into a fistula; it is therefore essential that whenever the spinal or cerebral dura is opened and left open, as occurs in all decompression operations, there must be a most careful closure of the overlying tissues. The muscles, fascia and subcutaneous tissue must be closed in separate layers. Cerebrospinal rhinorrhea may occur in association with pituitary diseases. The craniopharyngeal sinus is the embryonic remnant of Rathke's pouch from which the anterior lobe of the pituitary develops. At times this sinus remains patent and a copious intermittent discharge of cerebrospinal fluid may occur.

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* Anton, Gehirndruckentlastung mittels Balkenstiches, Deutsch. Zeitschr. f. Nervenh. Leipzig, xliii (1911–1912), p. 347.

Sinus Thrombosis.—It is usually said to be due to three causes; (1) Marasmus or general debility; (2) trauma; (3) infection. In very young children who have been undernourished and have had gastric or intestinal disturbances, thrombosis of the sinuses, particularly of the longitudinal sinus, has been observed. It is a difficult diagnosis to make and probably is not recognized because it is not suspected. Probably the symptomatology that Gordon Holmes has described in traumatic lesions of the longitudinal sinus occurs in these marantic cases (see below).

Traumatic nonseptic sinus thrombosis occurs with a fracture of the vault. Holmes saw many such cases in soldiers and pointed out wherein they differed from thrombosis of the cortical vessels. With the latter the motor centers lying nearest the Sylvian fissure, namely the face and arm centers, are most markedly involved, while in this type of thrombosis the leg center, and frequently both leg centers are most affected with only slight involvement of the arm and face. Traumatic cases without infection do best if left alone. The temporarily impaired circulation may be sufficiently restored to take care of the cerebral centers that have suffered injury from stasis of blood. If operation has to be resorted to, great care should be taken to promptly plug the sinus, preferably with a large piece of muscle, which is to be left in place permanently.

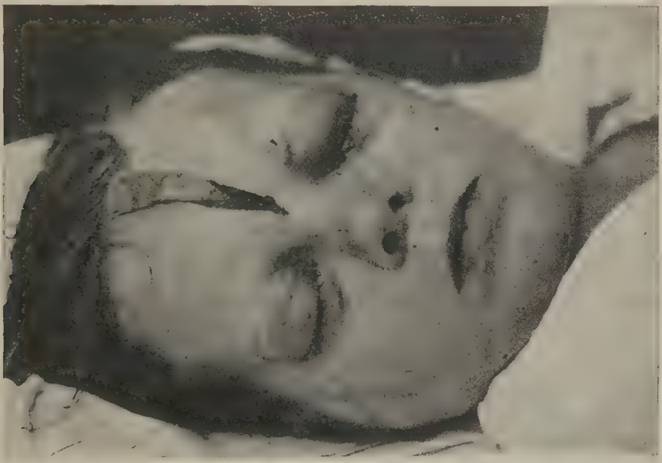

Fig. 10.—Cavernous sinus thrombosis following infection over the forehead. Note the large vessels in the lids. Incision over the forehead was an attempt to drain the primary infection.

Infective sinus thrombosis is justly one of the most dreaded of diseases for when it attacks certain sinuses the surgeon is absolutely helpless. An insignificant pustule about the orbit may cause thrombosis of a superficial vein and this thrombosis spread to the *cavernous sinus* and lead to pyemia and death in a few days. One can never tell when such an insignificant infection may have such an outcome, consequently proper measures in every infection are the sole prophylactic measure to prevent such a catastrophe. Incision of such infections is to be avoided until the abscesses is well walled off. At times an old chronic infection in the antrum sphenoid or ethmoid sinuses may be lighted up and give rise to a cavernous sinus thrombosis.

Cavernous sinus thrombosis is invariably fatal and the cases of reported recovery probably had some other condition (Fig. 10). Ballance* has advocated operating for these cases by evacuating the clot and draining the sinus. No case has ever survived and clearly the operation is wrongly conceived. If the clot could be completely evacuated the procedure might be considered but this is impossible in the case of the cavernous sinus.

The lateral or sigmoid sinus may be the seat of a thrombosis in association with a mastoid infection. In fact this is not so very uncommon. In these cases, if the diagnosis is made early, tying the jugular vein below the thrombus and laying open the entire vein and sinus, if necessary to the torcular, frequently saves the patient's life. At times in thrombosis of this sinus an abscess forms in the vein. Lateral sinus thrombosis may have associated with it a subdural (perisinuous) abscess, a meningitis or a true brain abscess. The symptoms of a lateral sinus thrombosis are quite characteristic. The patient has had an otitis media with mastoid tenderness and swelling. Suddenly a chill occurs followed by a septic temperature, headache, nausea and vomiting and great prostration; during the height of the fever the pulse is rapid and a high leukocytosis is usually found. The examination at this stage may show no tenderness or thickening of the jugular but a positive blood culture at this time clinches the diagnosis. Even though there be no other intracranial involvement the eye grounds may show some tortuosity of the vessels and early choked disc. Crowe claims that compressing the opposite jugular, if one is thrombosed, increases the tortuosity of the vessels and the choked disc. I have never been able to confirm this observation.

If the process has gone on for sometime intracranial symptoms may be more prominent due to the interference with the cerebral circulation. The symptoms of increased cerebral pressure described elsewhere may be present or local paralyses of the cranial nerves around the base of the brain may develop, hoarseness, dysphagia, dyspnea and slow pulse.

Infections of the longitudinal sinus are far rarer but in a large percentage of cases follow injury. A thrombosis may spread back from the nose and give the symptoms described by Holmes. Though not quite as hopeless as a cavernous sinus thrombosis because more accessible, it is doubtful if any case has ever been saved.

The principle laid down by Zanfel and practiced with such brilliant results by MacEwen† must be adhered to. The sinus must be laid open from one end of the clot to the other and drained.

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* Ballance, Chas., "Surgery of the brain and its membranes" (1907).

† MacEwen, W., "Pyogenic infective diseases of the brain." Glasgow, Maclehose (1893).

Arteriovenous Aneurysm.—This is invariably traumatic in origin and occurs as a result of a fracture. A communication develops between the cavernous sinus and the internal carotid artery or its single intracranial extradural branch, the ophthalmic artery. The condition usually develops in the course of a few days or weeks after the injury. The two most striking symptoms are an incessant bruit in the head, which the patient notices and which can be heard with a stethoscope, and a pulsating exophthalmos. The veins about the lids and in the conjunctiva may be greatly dilated. The vessels in the eye-ball may be seen to be dilated with the ophthalmoscope. Headaches are a frequent accompaniment of the condition.

The treatment consists in occlusion of the carotid artery. In order to avoid the danger of common carotid ligation it is well to have the patient practice compression of the carotid for a number of weeks prior to the operation so that cerebral anemia may not occur. The operation is best done under local anesthesia by placing a Matas* band around the vessel and compressing it until the carotid circulation is markedly impeded. If the patient nevertheless develops cerebral symptoms the band can be readily removed. A few cases of spontaneous cure are reported.

Inflammatory Processes of the Cerebral Membranes. Pachymeningitis Externa.—Extradural inflammatory processes are rare. They usually occur in association with some disease of the bone as osteomyelitis of pyogenic or syphilitic origin. If not dealt with early the process may have extended through the dura and produced a meningitis or localized abscess. Free drainage is the treatment indicated.

Pachymeningitis interna hemorrhagica is often traumatic in origin. It has also been described in children by Sutherland† in association with scurvy and whooping-cough. A blood clot lying between the dura and arachnoid becomes encapsulated and may give local pressure symptoms. It may produce general or focal pressure symptoms and may mislead the surgeon into thinking he is dealing with a tumor. The only case which I have seen followed a fracture and had such an extensive process that it was quite impossible to remove the membrane entirely. The brain was markedly compressed and the sudden relief of pressure apparently brought about the death of the patient.

Leptomeningitis, commonly called meningitis, may be acute or chronic. It may be produced by a variety of organisms and may be a primary or secondary disease. The infection may be a general infection of the meninges or the process may be localized and may then be in the nature of an abscess. The exudate may be thick and purulent or may be serous. Cultures occasionally are sterile which has given rise to the confusing term *meningismus.* This term is applied to those cases in which clinically the patient presents symptoms of meningitis but bacteriologically no organisms are found. Such cases, however, show histologically an inflammatory reaction and it would be far better if such cases were spoken of as serous meningitis. Recently Fammler has shown that these cases show an abnormal colloidal gold reaction which is corroborative evidence that this is a form of meningitis. The clinical pictures vary a good deal, depending upon the acuteness of the infection and also the type of organism.

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* Matas and Allen, Occlusion of large arteries with removable metallic bands, etc., Jour. A. M. A., lvi (1911), p. 233.

† Sutherland, G. A., On hematoma of the dura mater associated with scurvy in children, Brain, xvii, p. 27.

Cerebrospinal meningitis in its epidemic form, though at times it may be sporadic, is due to the Diplococcus intracellularis of Weichselbaum (a Gram-negative diplococcus). At times in young children the process is localized at the base and has been called by Barlow, Koplik* and others, posterior basic meningitis (Fig. 11). It is essentially the same disease and is not a separate clinical entity. Clinically these cases may become chronic and due to the obstruction to the cerebrospinal fluid develop hydrocephalus. The process is confined to the meninges and usually the encephalon shows no changes. When very acute this disease may be fulminating in character and lead to death in 24 hours. The onset may be insidious, headache, vomiting, irritability, and fever, followed rapidly by retraction of the head and heels which may become extreme. The temperature usually is high without remissions. The leukocyte count may exceed 100,000 though it usually does not get higher than 40,000 cells. During an epidemic various types may be recognized. (1) Those with marked cranial nerve involvement. (2) Those with general meningeal irritation but no focal symptoms. These cases may have convulsions especially in young children. Hyperesthesia and restlessness may be extreme and stroking the skin may bring out the "tache cerebrale" which is really nothing more than a skin reaction due to marked vasomotor sensitiveness. (3) A more chronic form may be observed with stupor, some fever, bulging fontanelle if the sutures have not united, choked disc and slow pulse—the picture of cerebral compression. (4) Abor-

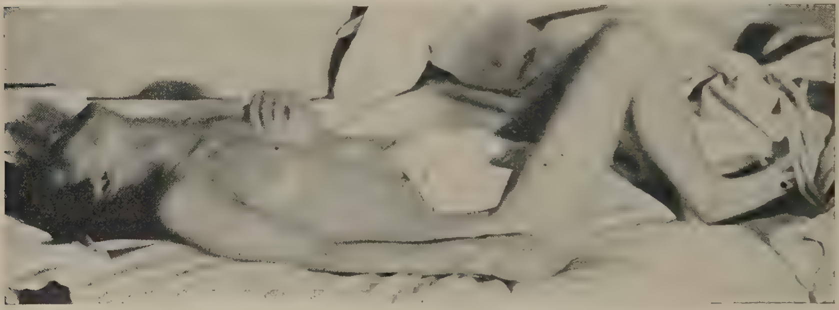

Fig. 11.—Epidemic cerebrospinal meningitis. Note the extreme retraction of the head.

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* Koplik, H., Posterior basic meningitis, Mt. Sinai Hospital Rep., N. Y., iv (1905), p. 105.

tive cases. These are seen particularly during an epidemic and show evidence of meningeal irritation with rapid subsidence of the symptoms.

The diagnosis is not difficult as certain signs are always present, namely, rigidity of the neck, fever, Kernig's sign— inability to extend the leg when the thigh is flexed to 90°—and Brudzinski's sign—flexion of the legs when attempt is made to flex the head. The finding of the intracellular diplococcus clinches the diagnosis. The cases at times show a macular or petechial eruption and the disease therefore has sometimes been spoken of as spotted fever. The treatment consists of the repeated intraspinous administration of Flexner's serum. This has reduced the mortality tremendously. Whereas before the use of serum the mortality was as high as 75 per cent, now in some epidemics it has been possible to reduce the mortality to 25 per cent and even less. The injections should be given every 6 to 12 hours until the fluid is clear and the organisms have disappeared. In spite of the administration of serum a certain number of cases develop into a chronic form. The same may occur if serum has not been given. In these cases the spinal fluid becomes clear, organisms disappear, but the child presents the picture of a hydrocephalus. These are the cases that have had the process primarily at the base and the exudate has become organized and causes an obstructive hydrocephalus. Still another type must be recognized. At times an acute case does very well for a few days and then the spinal fluid becomes so thick that it no longer runs through the needle or lumbar puncture yields a little fluid under slight pressure. In both of these instances the process may have become localized to one or both lateral ventricles practically producing an intraventricular abscess and if the fontanelles are not closed there may be very marked bulging. The intraventricular administration of serum in these cases has cleared up some of them.

Permanent drainage of the subarachnoid space in the cervical region is, I believe, undesirable nor do I believe that the puncture of the basal cisterna, as suggested recently by Ayer, is a safe procedure. The constant to and fro movement of the medulla during respiration makes such a puncture exceedingly dangerous. The danger of producing a Claude Bernard *piceur* is very great. More recently Ayer has published a large series of cases without any fatality so that in careful hands the procedure may be carried out.

The irrigation of the subarchnoid space by a combined spinal puncture and craniotomy has also been suggested recently again by Alford* but his experimental work along these lines is not very encouraging since his work has shown that all parts of the subarachnoid space cannot be reached in this way. It would seem that the irrigation of the subarachnoid space finds its counterpart in the flushing of the peritoneum for peritonitis that had great vogue some 15 years ago but was soon given up. We know now that the peritoneum can take care of the exudate far more effectively if it is not traumatized and that the essential point is to get rid of the primary focus of infection, namely, the diseased appendix. The problem seems to be a very similar one. Repeated lumbar puncture with withdrawal of cerebrospinal fluid as advocated by Quincke* has seemed to cure some of these cases.

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* Alford, L. B., Irrigation of subarachnoid space, Journ. of Nerv. and Ment. Dis., xlviii (1918), p. 359.

Meningitis of Pyogenic Origin.—A variety of organisms may be responsible for this. The condition may follow a trauma or infection elsewhere in the body. It is even more serious than when due to the Diplococcus of Weichselbaum. The symptoms are practically the same as have been described. If a focus of infection can be found its drainage may clear up the meningitis. The most favorable types are those following an otitis media and mastoiditis or a nasal infection.

The irrigation of the spinal meninges in these cases is equally ineffectual. Leighton has reported several apparently desperate cases that recovered following a drainage of the spinal meninges. I have tried this myself in several cases but without success. One must always ask oneself whether these cases though apparently desperate might not have recovered without surgical intervention.

Meningitis serosa, first so-called by Billroth but studied in great detail by Quincke, is characterized by an increase of spinal fluid which always remains clear. At times the cell count is increased but sometimes is normal.

The symptoms are those of a meningitis of moderate severity but pathologically the findings are often quite negative. The condition apparently is not due to any organism and cultures of the spinal fluid are negative. It may be due to toxins that get into the subarachnoid space and the process may be in the nature of a chemical meningitis. It is this condition probably that comes on after sun stroke or in association with alcohol. The condition known as *meningismus* probably belongs to this group. Lumbar puncture often relieves the condition. Cushing has decompressed some of these cases with marked relief.

Ependymitis.—Inflammatory processes may occasionally affect the ependymal lining of the ventricles as part of a meningeal infection. The process may be confined to the ventricles but I do not agree with Cushing that it is a specific disease. As a result of the location of the lesion in the ventricle, hydocephalic symptoms are a very prominent feature. The diagnosis may be impossible and lead to a suspicion of brain tumor. If the cause can be determined treatment along lines similar to those outlined under meningitis should be followed.

Tuberculous meningitis occurs most frequently in children. It is always fatal. The few recorded cases of recovery are open to question. The onset of the disease is usually insidious though at times it runs as acute a course as a septic meningitis.

The patients die apparently from the gradually increasing pressure symptoms. Internal hydrocephalus is a common accompaniment of the condition and these patients lapse into a semi-comatose state in which they may remain for days or weeks before they finally succumb. The differential diagnosis may be readily made by careful examination of the spinal fluid. Bernstein* was the first to show that careful painstaking examination of the spinal fluid showed tubercle bacilli in practically 100 per cent of cases. In addition, the lymphocytes are increased in the fluid and not infrequently tubercles may be found in the retina. Withdrawal of cerebrospinal fluid by lumbar puncture or ventricle puncture to control the pressure symptoms is the only treatment that has been suggested.

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* Quincke, Ueber meningitis serosa, Volkmann's Samml. Klin. vorträge, Leipzig, No. 67 (1893).

Syphilitic Meningitis.—The disease may follow two general types, either that in which the meningeal symptoms are the prominent or the sole ones, or the focal type in which the picture is that of a brain tumor and varies considerably depending on the location of the process.

In the meningeal type the patient may present the symptoms of a meningitis but rather chronic in character. For the details of the symptomatology and diagnosis, the reader is referred to Chapter CLVIII Vol. VI. We are here concerned with that phase of the subject which calls for surgical intervention. Choked disc is a common accompaniment of syphilitic meningitis. Some ophthalmologists still claim that the changes in the eye grounds in syphilis are different than those seen in tumor. I feel convinced that this is not so. I have on several occasions seen a choked disc called syphilitic which subsequently proved to be due to tumor. Furthermore, I believe these eye changes in syphilis are a pressure phenomenon just as is the choked disc due to tumor. An unrelieved syphilitic choked disc goes on to atrophy and blindness just as it does in brain tumor. The treatment therefore should be directed so as to afford prompt relief by decompression, but when to decompress is still a disputed question; Oppenheim† advised waiting for many months, Allan Starr for three months, while Horsley‡ advocated not waiting more than six weeks. Even this may be too long. The determining factors should be the amount of vision the patient has, the length of time the process has existed, and the ophthalmoscopic picture. If vision is very seriously impaired decompression should be done at once and then specific treatment instituted either with mercury or salvarsan. In fact, Horsley pointed out, and others have verified his observations, that specific therapy is more effective after the intracranial pressure has been relieved. Whether it is better to use salvarsan or mercury in these cases is still an open question. There have been bad results reported from the use of salvarsan in the presence of choked disc so that great care must be exercised. It cannot, however, be said to be contra-indicated.

In the cases of focal syphilitic lesions if the patient shows pressure symptoms they must be relieved; if these do not clear up promptly with medication the same operative procedures should be employed that are indicated for brain tumors. If the location of the lesion can be determined the process, often a gumma, should be dealt with as any other brain tumor. Horsley and Gowers in the early nineties expressed the view that many gummata could not be made to disappear with specific treatment and therefore should be operated. This idea has made very slow progress but is now being recognized. The headaches that are such a prominent symptom of both syphilitic and non-syphilitic intracranial lesions may be temporarily relieved by mercury or salvarsan and as a result valuable time is lost. The headache of syphilis alone may be an operative indication, though if there are no other pressure symptoms drug therapy should always first be tried.

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* Bernstein, E. P., The value of lumbar puncture with particular reference to the diagnosis of tuberculous meningitis, Med. News, N. Y., lxxxvi (1905), p. 1105.

† Oppenheim, H., "Die Geschwülste des Gehirns." Wien (1902).

‡ Horsley, V., The surgical *vs.* the expectant treatment of intracranial tumors. Address before German Society of Neurologists, Berlin (Oct. 6–8, 1910).

Lesions of Blood=vessels.—Intracranial Hemorrhage.—One of the common causes of acute cerebral compression is intracranial hemorrhage. The condition as it occurs at birth has been discussed elsewhere as it was desirable to group all forms of birth injuries together. Furthermore, as pointed out at that place, the picture and diagnosis of hemorrhage in the newborn is so different from that which occurs in children whose nervous system has developed completely, that a separate consideration has been deemed quite in order.

Intracranial hemorrhages may be grouped under four headings:

1. Extradural.
2. Subdural.
3. Subarachnoid.
4. Intracerebral.

Extradural.—The dura in children is very firmly adherent to the bone; the younger the child the more marked is this the case, but a moderate pressure will peel it away and permit a considerable collection of fluid. Extravasation of blood in the extradural space is practically always a result of trauma and usually associated with a fracture. The blood as a rule is arterial in origin, though it may be venous. Venous oozing is seen at times following osteoplastic operations and may be a cause of alarming symptoms. The middle meningeal artery supplies blood to the dura. It divides into two branches, the anterior and posterior, and may be injured anywhere in its course. The most common site of injury is in those few millimeters of its length when it passes from the foramen spinosum to the dura. At this point it is unsupported and therefore is readily torn by a sudden jolt of the brain and membranes as occurs in a fracture. The blood collects in a small pocket which it makes by peeling off the dura from the bone and gradually peels off more and more dura as the hemorrhage progresses and may make a tumor of considerable size. Such a clot may vary in thickness from a millimeter or two to a centimeter. The thickest part of the clot is usually at the point of rupture of the vessel and this point will give rise to the most marked focal symptoms unless perchance it lies over a silent area. Careful studies have been made to determine the points where such extravasations occur most frequently with a view to determine the site of operation. The minute directions for locating these hemorrhages have now little more than historical interest for, in the first place, surgeons no longer should undertake any work through a trephine opening, and secondly, the site for an operation should be determined by a study of the neurological symptoms.

Hemorrhage from the middle meningeal artery gives rise to a very characteristic clinical picture. In the typical case the patient may become momentarily unconscious as a result of a blow on the head and then regain consciousness and appear perfectly normal. After a time interval varying from a few minutes to several hours, the patient passes through the various stages described under compression: headache, drowsiness, finally unconsciousness, and, unless prompt relief is afforded, goes into deep coma as a result of the rapidly increasing compression. The period of consciousness between the two periods of unconsciousness is spoken of as the *lucid interval* and is a picture that has been recognized for a long time. In a middle meningeal hemorrhage, the various changes in blood pressure seen in acute cerebral compression are more strikingly observed than in any other type of case. In such an injury the preliminary period of unconsciousness may be absent and before the general compression symptoms develop focal ones may be noted. These first are irritative in character for moderate pressure on the cortex or any nervous tissue first irritates before it paralyses. Since the hemorrhage of a middle meningeal lesion always occurs in the middle fossa the symptoms frequently arise from irritation of the motor cortex, and since the blood wells up from the under surface of the brain the most dependent portion of the motor area will be affected, the face center, and then the hand, the arm, and ultimately the leg. Convulsive movements may first be noted followed later by paresis or complete paralysis. With the increase of the symptoms the general symptoms of compression may come on and obscure the focal character of the picture. In right-handed individuals a hemorrhage on the left side may produce a motor aphasia. In children, particularly, the point of injury may be over the occipital lobe as they far more frequently fall on the back of the head as in ice and roller skating. In such cases, if the child is old enough to describe its symptoms, hemianopsias may be noted. If the hemorrhage is far forward, due to injury of the anterior branch of the meningeal artery, in addition to motor aphasic symptoms conjugate deviation of the eyes may be noted. At first while there is mere irritation of this center which lies in front of the face and hand center, this conjugate deviation is toward the side opposite the lesion, but as paralysis sets in the eyes are turned toward the side of the lesion. Changes in the eye grounds occur as the pressure increases, congestion of the vessels, tortuosity, and ultimately edema about the disc.

If there are focal symptoms, the side on which the hemorrhage has occurred is readily recognized, but frequently it may be difficult to determine the side on which the hemorrhage has taken place. This difficulty may be due to the absence of any focal symptoms or because they are masked by the general symptoms. By the time the patient is seen, the general symptoms may predominate, but there may be a history of focal symptoms. A very careful neurological physical examination is always essential. A single pathological reflex may give the surgeon the necessary clue. One point cannot be emphasized too strongly; *the location of the hemorrhage should be determined by the neurological symptoms and not by the location of the blow on the skull* for not infrequently the lesion has been a contrecoup one so that the lesion is on the side opposite to that of the blow. It should be the invariable rule that in all cases of suspected intracranial injury frequent blood pressure observations be taken. In fact the surgeon who neglects this procedure is guilty of a grave offense.

Treatment.—Stopping the hemorrhage and evacuating the clot are the two objects aimed at. One of two operative methods is indicated, either to turn down an osteoplastic flap or to approach the middle meningeal through a subtemporal incision similar to that used for a subtemporal decompression. The advantage of this latter procedure is that it is less formidable and less time consuming and can readily be carried out on both sides in those cases in which the side of the lesion is in doubt. It occasionally happens that one bleeding point cannot be found because there have been numerous small bleeding points or the acute hemorrhage has stopped. In such cases evacuating the clot may be all that is required or it may be necessary to insert a small cigarette drain (gauze covered with gutta percha tissue). Such a drain must be removed in 24 hours and never replaced. It is far better, however, to make a painstaking toilet of the field than to leave a drain, on account of the great danger of an infection.

Subdural Hemorrhage.—This type of hemorrhage occurs more frequently in association with contusion and laceration. It may also occur as the result of a very slight trauma of which the following case is a striking example.

A young girl who had a good family history, no evidence of syphilis, or hereditary illness of any sort, suddenly sneezed and within a few minutes, without loss of consciousness, developed a homonymous hemianopsia. After several years she had a few attacks of focal epilepsy which never recurred. Her hemianopsia first changed to a quadrantic hemianopsia and finally completely disappeared.

The symptoms and clinical course may be identical with what has just been said of extradural hemorrhage. Blood in the spinal fluid will help to identify the condition since a subdural hemorrhage has almost inevitably associated with it enough subarachnoid injury to enable blood to get into the subarachnoid space. As the inner surfaces of the dura and arachnoid are not at all attached to each other as is the dura to the bone, blood does not collect at one point but spreads over an entire hemisphere or even into the posterior fossa and along the cord. If there is a great deal of contusion a hematoma may develop at such a point and then produce focal symptoms. Evacuation of the clot is carried out as in the extradural cases. In addition, badly lacerated brain tissue should also be removed either by excision or by irrigation with warm saline solution or by suction with a catheter and syringe. This tissue should be removed as it forms a scar which later on may be the cause of a Jacksonian epilepsy. In any operative interference for subdural hemorrhage, it must be kept in mind that intradural pressure is increased and that before opening the dura in such a case the tension of the dura should first be carefully observed and if the pressure is greater than normal it must first be reduced by removal of cerebrospinal fluid either by lumbar or ventricle puncture. It is well in this type of case to leave the dura open, in other words to do a typical subtemporal decompression, in order to make room for the edema which always follows any extensive manipulation of the brain.

Subarachnoid hemorrhage might almost be called a condition peculiar to childhood. When due to trauma it, of course, is no different than in the adult. The blood infiltrates the pia arachnoid and may obscure the surface of the convolutions. On account of the peculiar structure of the arachnoid, the hemorrhage never forms a very thick clot but rather a thin film. It can readily be evacuated by numerous punctures of the arachnoid with a fine needle. There is no symptomatology that is characteristic of this condition; it is usually found in association with contusion and laceration. The blood in the arachnoid should be evacuated, though as a rule such a film of blood is absorbed. I have seen a cortex which was completely obscured by a subarachnoid hemorrhage look perfectly normal after four weeks. Occasionally these exudates are not absorbed and there remains a collection of clear or slightly brownish stained fluid. This condition has been variously described as a *subarachnoid cyst*, *arachnoiditis*, *chronic serous meningitis* or *pachymeningitis*, and with this there may be degeneration of the cortex. A large variety of symptoms may be produced in these cases: All grades of mental impairment from the backward child to the low grade imbecile; there may also be epilepsies more frequently general than focal. Not a few of these children fall into the criminal class. The evacuation of these cysts results in some cases in improvement unless there are secondary changes in the cortex but where there is cortical degeneration the evacuation of the fluid is of little value. The condition of the cortex cannot be determined before operation so that exploration is necessary to determine the pathological status.

The large group of subarachnoid hemorrhages associated with the infectious diseases, particularly whooping-cough, measles, and scarlet, offer a further problem. Frequently the hemorrhagic exudate has an inflammatory character. Patients with this condition usually have one or several convulsions, often focal in character, and this may be followed by a paralysis. The last word has certainly not been said on this subject but in view of the fact that the condition occurs in the course of an infectious disease and that these subarachnoid exudates usually are absorbed, it seems better not to operate upon these conditions. The sole exception is the rare case of hemorrhage associated with definite focal paralytic symptoms that develops suddenly after a violent paroxysm of coughing during whooping-cough. In these cases a bone flap should be turned down and the clot evacuated.

Intracerebral hemorrhage can be disposed of in a word. It has no place here as it is associated with arteriosclerosis and periarteritis and consequently a disease of middle and old age.

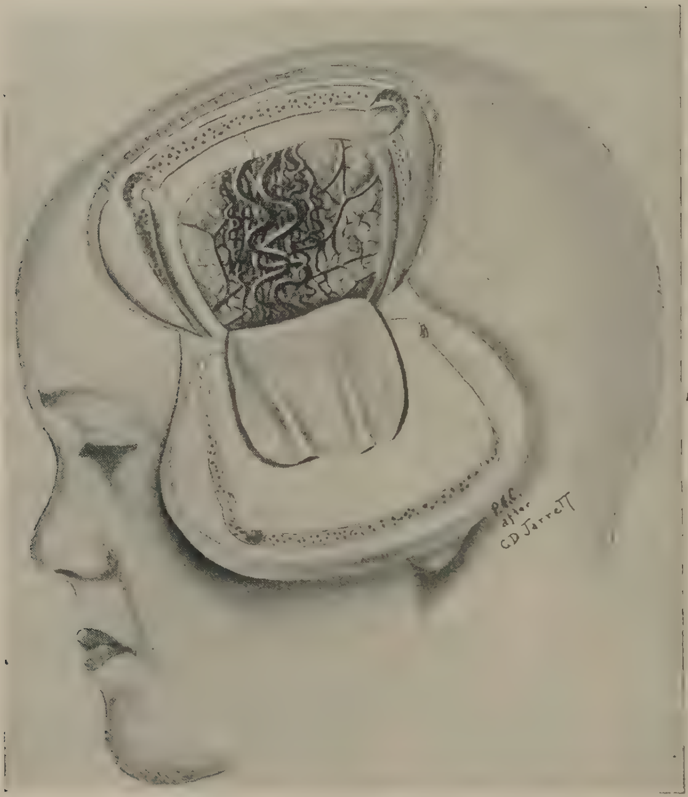

FIG. 12.—Intracranial telangiectasis, cortical type.

One or two cases have been reported by Marriott of arteriosclerotic changes in a child in which a capsular hemorrhage occurred but this is such a very extraordinary occurrence that it need not be considered, especially as the advisability of any operative interference directed toward relief of such a hemorrhage is very doubtful. Even in older people with hemorrhage from arterial disease it is by no means an accepted procedure. In children when such arterial disease exists it seems equally inadvisable.

Intracranial Telangiectasis (Figs. 12–14).—This condition has been at times called a blood vessel tumor but in view of the pathology of these cases it seems certain that the process has none of the ear marks of a new growth but is a congenital anomaly. The condition is frequently associated with nevi on the face or head. The process may be confined to the vessels of the dura or to the vessels lying in the pia arachnoid or may affect both. The great importance of the condition is that it is one of the causes of Jacksonian epilepsy in children. In the absence of syphilis a Jacksonian epilepsy in a child may be due to this condition. The differentiation of this condition from tumor is at times quite impossible. The author some years ago reported two cases of this sort and summed up the points in the differential diagnosis.

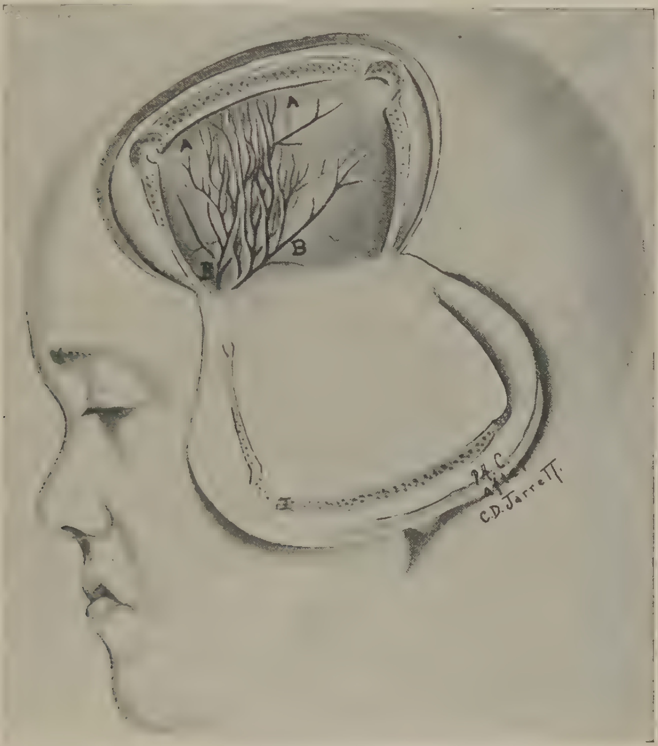

Fig. 13.—Intracranial telangiectasis, dural type.

These were:

1. Attacks of Jacksonian epilepsy occurring at long intervals in non-syphilitic young persons.
2. Unconsciousness of long duration.
3. No evidence of increased intracranial pressure.
4. A very slow progression of symptoms.
5. Telangiectases on the head or face.

Aneurysm.—This condition may be said never to occur in children and therefore has no place here. It is associated with arterial disease and may give symptoms identical with tumor. A closely allied condition, however, *arteriovenous aneurysm*, does occur. It is always associated with a trauma and usually develops at the base of the brain either around the internal carotid or vertebral arteries. It may give rise to a variety of pressure symptoms depending on its location. The symptoms may vary somewhat from day to day due to the changing pressure conditions. They progress, however, extremely slowly and may be stationary for months or even years, therein differing from a tumor. Ophthalmoscopic examination sometimes helps in the diagnosis. In a case of the author's, the location of the arterio-venous aneurysm was suspected by the enlargement of the arteries in the fundus. The traumatic aneurysm was of the ophthalmic artery and the symptoms arising from it were controlled by partial occlusion with a Matas band. The most annoying and distressing symptom is the constant roaring in the head night and day, which makes these patients very miserable; at times this is so annoying that it leads to suicide. If the internal carotid is affected, occlusion by ligation may alleviate the symptoms; if it has to be tied, digital compression should first be practiced for several weeks, the period of compression being constantly lengthened, and then when the patient shows no signs of cerebral disturbance due to the transient anemia, the carotid should be partly occluded by means of a Matas or Halsted

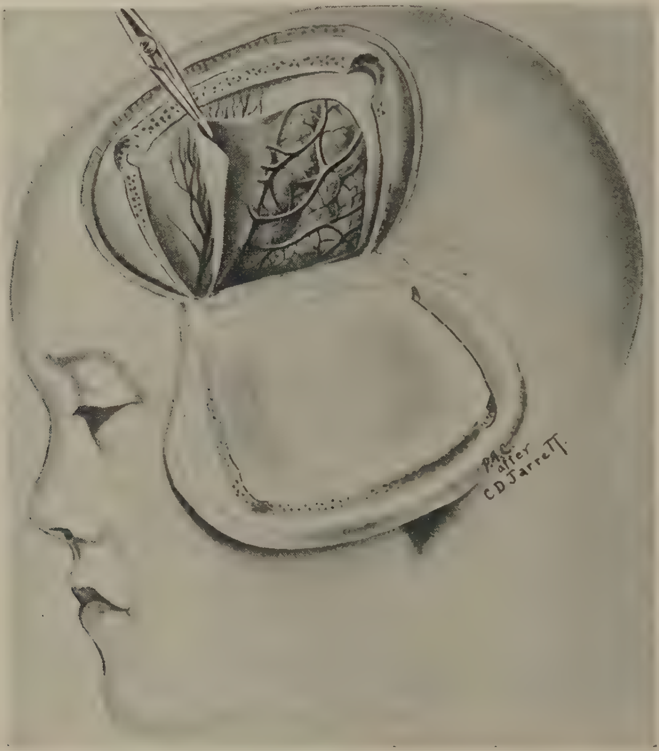

Fig. 14.—Intracranial telangiectasis, dural type.

metal band. The purpose of the digital compression is to develop a collateral circulation.

Thrombosis and Embolism.—Arterial thrombosis, like aneurysm, is not a disease of childhood. It occurs in association with degenerative processes in the vessels, arteriosclerosis and endarteritis. Even in adults it never is amenable to surgical measures. Septic thrombi have been discussed elsewhere under infections of the meninges.

Embolism occurs in association with septic or chronic endocarditis. The area whose blood supply is cut off may be taken care of by collateral circulation and the symptoms arising from the temporary anemia of the region may clear up completely. If the vessel affected is of considerable size, the collateral circulation will not take care of the affected area and an area of softening develops. For this no surgical measures avail. There may result, however, spastic conditions which may require orthopedic measures to relieve the contractures. Of these measures, tendon lengthenings or tendon transplantations or muscle group isolation as described by Schwab and Allison* are the most effective.

Inflammations of Brain.—There are a number of inflammatory conditions of the brain which do not require surgical treatment, yet must be mentioned here because occasionally the pediatrician calls in a neurological surgeon to see them and give his advice on certain phases. These conditions are acute *poliomyelitis* and *encephalitis lethargica.* A full discussion of the diseases will be found elsewhere. At times both of these conditions may produce the symptoms of an acute meningeal infection, vomiting, headache, stupor or somnolence, fever, rapid pulse, choked disc, and, in the late stages, a slow pulse, and respiratory disturbances. Paralyses or focal irritative phenomena (focal epilepsy) may also be present. The question may then be whether the lesion is one that may be relieved by operation. If the diagnosis of poliomyelitis or encephalitis has been made, the wiser course is not to interfere surgically as the process is so diffuse that the mere relief of pressure is of little avail. Such cases have occasionally been operated upon and have recovered. It seems probable to me that the operation was not a factor in the outcome.

Brain abscess (Fig. 15) is not an uncommon disease in childhood. The signs and symptoms that should make one suspect the condition and the possibilities in the way of treatment are so little known that the cases frequently are not discovered until autopsy. Of 223 cases that Gowers collected, 72 occurred in childhood. An abscess may develop in the brain substance as a result of *trauma with direct infection, trauma with secondary infection* via the blood stream, *pyemia* in which case the abscesses usually are multiple, and *direct extension from an osteomyelitis* of the bone. The most common origin for such an extension from the bone is either following mastoid disease or an infection of the nasal sinuses. What the underlying factor is that predisposes brain tissue to infection has not been proven. Certain it is that whereas meninges are readily infected, brain tissue is not. I have had two of my students busy themselves with this problem but thus far they have been unable to produce a brain abscess experimentally. In young children the abscess may develop from an otitis media without any apparent involvement of the mastoid since the mastoid cells during the first year or two of life are poorly developed. The infection may pass directly by thrombosed veins through the dura into the cerebral substance.

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* Allison, N. and Schwab, S. I., The result of muscle group isolation in the treatment of paralysis of the extremities, Amer. Orth. Surg., ix (1912), p. 224.

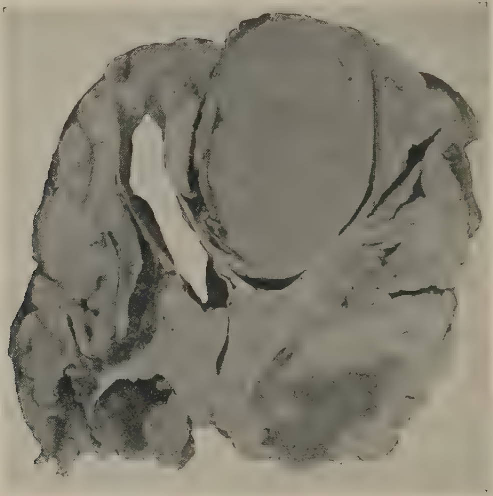

Fig. 15.—Brain abscess produced by birth injury causing an obstructive hydrocephalus. Child lived 2½ years.

Pathology.—A variety of organisms may be found. In addition to the pyogenic organisms, pneumococcus, typhoid, influenza and pyocyaneus have been isolated. The abscess at an early stage may be merely a broken down ill-defined area of brain substance containing pinkish necrotic material. The longer it has been present, however, the thicker is its surrounding wall and this in course of time becomes so rigid that it may offer a serious difficulty in the cure of the condition. Frequently the portal of entry is obscure or not discoverable and the patient presents the picture of an intracranial lesion without apparent etiological factor.

Symptomatology and Diagnosis.—One of the most striking features of brain abscesses is the prolonged latent period during which no trouble is apparent, neither fever, leukocytosis nor any of the symptoms that characterize infections elsewhere in the body. There may be the history of an acute stage to the condition when the abscess first developed, fever, chills, headache and vomiting, but far more frequently there is not only no such history obtainable but the entire past history is negative.

MacEwen divides the disease into three stages: (1) The initial stage. (2) The stage in which the abscess is well developed. (3) The terminal stage. More frequently there are but two stages: (1) The latent period. (2) The terminal one. The insidious character of these cases is well illustrated by the one I have recorded in the section on hydrocephalus; this abscess was present from birth, was not suspected and only found at autopsy. Another instance occurred in a case of mine suffering with yaws. This boy had his abscess unquestionably for one and a half years, went to school, took part in athletics and stood very well in his school work and did not complain in any way. The symptoms may be focal in character depending upon the region that is involved or there may be only general pressure symptoms. A *slow pulse* is one of the most characteristic signs and combined with this in the late stages there may be other evidences of medullary compression as changes in the respiration (Cheyne-Stokes). Sudden stupor or delirium may be the first signs in other cases. An abscess may develop in any portion of the brain but the most frequent sites are the temporal lobe or cerebellum as a result of a mastoid infection or the frontal lobe as a result of an infection of the nasal sinuses.

Lumbar puncture, as Strauss has shown, often shows an increase in the polynuclear elements without organisms. This is apparently a reaction on the part of the cerebrospinal fluid when the abscess lies near the ventricle.

The prognosis is always very grave but by no means hopeless. The treatment should always be surgical. The same principles apply as with abscesses elsewhere in the body but certain precautions are necessary due to the peculiar intracranial conditions. Such a brain is always under increased tension so that when the dura is opened the brain bulges, but as soon as the abscess is incised the brain tends to fall away from the dura, and this gives an opportunity for pus to get into the subdural and subarachnoid spaces and produce a meningitis. Every possible effort therefore must be made to wall off these spaces.

The method of drainage offers a further difficulty. MacEwen appreciated this and used decalcified bone tubes. Certainly the ordinary rubber tube still used by many surgeons is harmful since its weight and rigidity causes further damage to the brain. Others have recommended tubes of wire springs but these have the same disadvantage. Barany* during the war found that gutta percha strips were by far the best. Care must be exercised to replace the drain into the bottom of the cavity if it is removed. The brain tissue is so soft that there is grave danger of making a false channel. To guard against this, MacEwen advocated never removing a drain once it had been inserted. Recently Coleman and Dowman† have reported good results by the slow evacuation of abscesses.

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* Barany, R., Die offene und geschlossene Behandlung der Schussverletzungen des Gehirns, Beiträge zur Klin. Chirurg. Tubingen, xcvii, Hft. ii (1915), p. 397.

† Dowman, Charles E., The treatment of brain abscess by the induction of protective adhesions between the brain cortex and the dura before the establishment of drainage, Archives of Surgery, vi (May, 1923), pp. 747–754.

Concussion, Contusion and Laceration.—These two conditions are so closely associated and so frequently pass one into the other that they have to be discussed together. There is no symptom or group of symptoms that may be said to be characteristic of either concussion or contusion and laceration, and with either condition symptoms of compression may develop.

Concussion and contusion are always due to some trauma while compression may be brought about by many other causes, tumors, inflammation or systemic diseases which may produce cerebral edema, as measles, scarlet or whooping-cough. It also may occur in association with nephritis or any condition that produces convulsions.

The very mildest picture of concussion one sees is that of the individual who receives a blow on the head and is momentarily unconscious. As soon as he recovers consciousness, he may have some headache, and feel dizzy and weak. After a few minutes or an hour he feels quite himself again. Such very mild cases invariably pass through a short period during which their pulse is slow. Sometimes as low as 50. This period is very transient and may not last more than half an hour. From this very mild type there are all gradations. The period of unconsciousness may be longer, the subsequent headache very severe, at times lasting for days, weeks or months. The dizziness may persist for a long time. At times this dizziness is brought on when the patient sits up in bed, at other times, when the patient suddenly bends over. There are numerous other symptoms that are encountered during or after the period of unconsciousness. There may be nausea and vomiting, respiratory disturbances, irregularity of respiration or periods of apnoea or there may be typical Cheyne-Stokes or Biot breathing. In other cases, general or focal convulsions are observed, loss of memory for the events immediately preceding the injury and then again this memory lapse may cover a number of hours preceding the injury. Memory usually returns in quite a characteristic way. The incidents farthest removed from the accident return first and gradually in an ever narrowing circle until finally the facts immediately preceding the accident are recalled.

The following case is a striking example of this. A man was driving an automobile over a street car track when a rapidly approaching car struck the machine and threw him out. He began to regain consciousness after about one hour; all memory for events preceding the accident for a period of several hours was lost. After a week he remembered he was out driving an automobile, after a few days more he recalled where he had been driving, and recalled approaching the street where the accident occurred. It was only many weeks after the accident that the patient remembered the moment preceding the accident when he was crossing the track and saw the onrushing street car.

There may be still other symptoms. A period of depression or attacks of restlessness or irritability, so severe at times as to be maniacal in character, and requiring that the patient be restrained. Finally, a large variety of paralytic symptoms due to injury of some cerebral centers. Of these, perhaps the most common are the ocular ones. a difference in size of the pupils or conjugate deviation of the eyes.

The question now arises, what is to be called concussion and what contusion and laceration and wherein does their pathology differ. There are two general views held on this matter, the one first advanced by V. Bergmann* that there is no demonstrable lesion in cases of concussion, and the view advanced by Kocher† that in concussion we have multiple, minute hemorrhages while in contusion we have larger extravasations. It must be apparent that since it is never possible to say in a given case that it is a pure case of concussion the view that Kocher advanced is hardly tenable. How can the pathology of a condition be worked out if the condition cannot be recognized clinically? My own preference has been to restrict the term concussion far more than has hitherto been the custom and to apply it only to the cases of momentary loss of consciousness with a temporary slowing of the pulse, a true *commotio cerebri*. I myself believe that no case of pure concussion ever dies and if we accept this definition of concussion, obviously no pathology for the disease can be worked out. All the other cases I consider varying degrees of contusion and laceration. At any time in the course of such a case, symptoms of compression may develop. It may be due to the edema of the brain which develops secondarily to the trauma, it may be due to hemorrhage or to a collection of cerebrospinal fluid which has become encysted by some aseptic inflammatory adhesions.

The age of the patient may influence the picture. What has been said applies to a child of any age. The younger the child, however, the more unstable the nervous system and the less developed are the inhibitory influences. Therefore, convulsions and varying degrees of irritability may be expected more frequently in young children. The same amount of injury will produce more irritation in the young child than in an adult. The brain in a young child also becomes edematous far more readily than in an older one so that compression occurs more readily. On the other hand, however, the distensibility of the skull in these young children more readily makes room for changes in volume. Nature has adjusted conditions wisely in still another respect. The processes of repair are much more active in younger individuals so that absorption takes place more rapidly.

Treatment.—No matter how mild the case is, the patient should always be kept in bed with the head slightly elevated. The time honored custom of putting an ice-cap or helmet on the head may be used though I do not see how it can be of any value. If the cold is not annoying to the patient it may be used. In very young children it may lower the body temperature and make the patient more susceptible to pulmonary infection especially if the period of unconsciousness is a long one. I have abandoned the ice-bag in this condition many years ago. Care of the bladder and rectum are very important. Retention of urine is a common occurrence in severe injuries of this sort. Catheterization should, however, not be resorted to until the patient has had ample opportunity to void. If the bladder on percussion is not too greatly distended it is quite safe to wait as long as 24 hours. The bowels should be kept open with enemas or mild cathartics. Violent purging should be avoided since the associated dehydration affects these children a great deal. The skin should receive great attention to avoid any bed sores and more than the usual care should be exercised in the application of external heat.

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* V. Bergmann, "Chirurgische Behandlung von Gehirn krankheiten." Berlin (1899).

† Kocher, T., "Hirnerschütterung, Hirndruck und chirurgische Eingriffe." Wien (1901).

If the period of unconsciousness is prolonged, lumbar puncture or ventricle puncture may be performed. On account of the danger of a respiratory collapse, if there is any evidence of increased intracranial pressure, ventricle puncture, is preferable to lumbar puncture. I have seen at least one case where a lumbar puncture with withdrawal of but a few drops of cerebrospinal fluid resulted in a respiratory collapse and almost instant death. This is a danger that has not been sufficiently emphasized.

Compression.—The cranial cavity is a rigid walled box, practically completely filled with a solid tissue. The only fluids in the cavity are the cerebrospinal fluid contained in the ventricles and the blood in the vessels. If for any reason the brain increases in size, as in edema, or something other than the brain, as a tumor or abscess, clamors for room in the cranial cavity, the only ways in which added space can be provided are by forcing out the fluids or enlarging the skull and in rare instances forcing out the brain itself through a hernial opening. The ease with which cerebrospinal fluid can be forced in and out of the cranial cavity is nature's method of taking care of moderate variations in intracranial pressure that may occur, but this provision is quite inadequate when great pathological increases in pressure occur. Then pressure is exerted on the blood-vessels and this is the prime factor in the production of compression. Compression may be of two types, acute and chronic. The acute condition may develop with edema, hemorrhage, or venous congestion associated with thrombosis while the chronic type is the result of tumors, abscesses or collections of fluid that gather slowly. If the brain is not displaced by a tumor but merely replaced by one, as in certain types of infiltrating glioma, there may be no compression symptoms that are demonstrable.

To understand the phenomena that are characteristic of compression the acute form has to be considered since that alone is the type that can be satisfactorily reproduced experimentally.

It will be remembered that the cerebral vessels have no vasomotor nerves and that consequently the cranial vessels are comparable to rigid tubes and the amount of blood in the cranial vessels at any moment is always the same. Therefore, to get more blood to a given region, the only mechanism available is to increase the rate of blood flow.

When there is any increase in intracranial pressure the blood-vessels become compressed, which leads to an anemia. Anemia has been shown to act as a stimulant to the centers in the medulla which control the circulation, consequently since the vessels are rigid walled to overcome this anemia, the only available means to combat it is to raise the blood pressure. If the intracranial pressure is increasing, as in a progressive hemorrhage, this heightened blood pressure only temporarily relieves the condition, for anemia again is produced and again the blood pressure is raised to overcome it and thus the blood pressure rises step by step until ultimately the medullary centers become exhausted and fail to react and then death rapidly follows. In such a death the heart continues to beat for some time after respiration has ceased. It is quite possible to divide the entire compression picture into a series of distinct stages, each of which has a fairly typical group of symptoms as suggested by Kocher. The first one is that of compensation. During this period the increased pressure may only result in forcing cerebrospinal fluid out of the cranial cavity, or there may be enough pressure to compress the veins of the scalp slightly and cause their dilatation. Patients in this stage may have headache and feel dizzy but usually show very few symptoms. If the intracranial pressure is enough to affect the circulation, the dilatation of the superficial veins of the head may be much more marked, and in young children they may stand out very prominently. The veins on the eyelids are sometimes the first to show this congestion and there may even be transient periods of cyanosis. The ophthalmoscope reveals in the eye grounds at this time fullness of the veins and tortuosity. In this stage the blood pressure is rising, and the vasomotor centers in the medulla are being called upon for help to keep up the compensation. Other symptoms are manifest; the headaches are severe, nausea and vomiting may be present, the mental processes may be dulled, but the patient may be readily roused, even to answering questions correctly though he immediately lapses into a drowsy state, akin to stupor. The respirations may show variations in depth and rhythm and become typically Cheyne-Stokes in character. The pulse may show marked fluctuations and become very slow, dropping as low as 45 or 50. From this condition a further stage of compression may develop in which the blood pressure is very high and the compensatory efforts of the vasomotor centers are taxed to their utmost. The cyanosis is marked, and all the symptoms above described are aggravated. The patient is unconscious and in a deep stupor from which he cannot be roused, his respirations are stertorous and then followed by periods of apnea. The eye ground changes are more marked and edema appears about the edges of the optic disc. Reflexes are often markedly reduced or even abolished. Bladder and rectal control may be lost and there may be paralysis of some or all extremities. At times there may be great restlessness with purposeless movements and violent thrashing around accompanied by mutterings. A patient may remain in this precarious stage for a long time and gradually recover, but it is readily understood that at any moment the medullary centers may become exhausted and the patient die. The longer the patient lives the better his chances of surviving the compression symptoms; however, a long period of stupor is so frequently accompanied by pulmonary symptoms that this becomes, after the first day, one of the gravest dangers.

Is the picture of acute compression seen in young children different from that in adults? The differences are not very great. The pulse shows greater variations, instead of being consistently slow it is very slow at one moment and then suddenly becomes very rapid. The stertorous breathing that is such a characteristic phenomenon in adults is rarely observed in children. The margin of safety in the third stage of compression is much smaller than in adults. The factors that make the picture of chronic compression in children different from that seen in adults are thin skull and readily separated sutures.

Treatment.—The problem is to relieve the rapidly increasing intracranial pressure. It is impossible to lay down hard and fast rules as to the proper procedure in each given case. A careful study of each case is essential and sometimes a period of observation is necessary to determine whether the intracranial pressure is going up or down. Then and only then can intelligent treatment be instituted. The extreme view that every case of increased pressure should be operated is as dangerous counsel as the view that operation should be avoided, because the mortality after operation in some hands has been as great as without operation. In some cases withdrawing cerebrospinal fluid may first be tried. This had always better be removed by ventricular puncture than by lumbar puncture, on account of the danger of a medullary collapse. If this procedure does not lighten the depth of the stupor and the patient's symptoms are not promptly alleviated, a decompression operation should be performed without delay. If the patient is comatose, no anesthetic is needed at all, though it is well always to have an anesthetist available as sometimes a patient begins to regain consciousness as soon as the dura is opened and the pressure is relieved. The operation can be carried out painlessly under local anesthesia and this is the ideal thing in all cases. To carry out local anesthesia satisfactorily, however, the coöperation of the patient is essential and this is rarely possible with very young children. I have, however, used local anesthesia several times on children between the ages of 12 to 14 with very satisfactory results.

In discussing the treatment of compression, only the procedures called for by the increased pressure have been considered. In many cases there are focal symptoms which require further surgical procedures or a modification of what is indicated by the compression alone. Such questions will be dealt with elsewhere and more properly belong under the treatment of focal lesions.

Brain Tumors.—These are not rare in children nor should they any longer be considered hopeless. The pediatrician must be readier than formerly to suspect the condition or think of it as a possibility. When a child presents any of the symptoms of general pressure recorded below, the physician should promptly study the case from the point of view of a possible intracranial lesion. He should not, as has happened too often in the past, think of this after weeks or months of aimless therapeutic endeavor. The addition of an ophthalmoscope to every physician's armamentarium should become a necessity.

Introductory.—Before discussing the various pathologic conditions under this heading, it is necessary to consider certain factors that are essential for the proper study of these cases from a surgical standpoint. The problem of the neurologic surgeon is somewhat different from that of the neurologist. The former, in addition to determining the underlying pathology of a given case, must be vitally concerned in the anatomic location of the lesion since he must know where to operate. For this reason I have found it very useful to take the history and make the physical examination along anatomic lines. The following outline has been found of great value.

NEUROLOGIC CASES

These points are to be inquired for besides the regular routine history and examination.

F. H.—Hereditary diseases of nervous system. Other nervous diseases (epilepsy, convulsions, paralysis). Similar diseases in family.

P. H.—Convulsions, (fits). Other nervous disorders.

In case of children, whether first or later child, and which on type of labor (instrumental and difficult; also if baby was asphyxiated).

P. I.—General account with special reference to sequence of development of symptoms. Memory, speech, general shape of head, *right- or left-handed*, etc.

Head.—Cranial Nerve Symptoms:

1. Loss of smell.
2. Dimness of vision, transient blindness.
3, 4, 6. Paralysis or weakness of muscles, double vision (if present, relation to two objects, blind spots).
5. *Sensory.*—Changes in face and cornea. Taste.
5. *Motor.*—Muscles of mastication.
7. Paralysis.
8. Hearing, dizziness and vertigo.
9, 10. Cardiac or respiratory symptoms, and loss of taste.
11. Trapezius or sternomastoid involvement.
12. Movements of tongue.

Subjective Sensations:

(*A*) General sensation, referable to extremities or face, tingling, etc.
(*B*) Vision (relation of persons or objects about them).
(*C*) Hearing.
(*D*) Taste.

NEUROLOGIC CASES.—(*Continued*)

- (*E*) Smell.
- (*F*) Dreamy state.

Upper Extremities:
- Loss of power or partial paralysis.
- Changes in sensation.
- Tremors, changes in nails, or unusual perspiring.

Abdomen:
- Bladder or rectal symptoms.
- Paradoxical incontinence, retention, or true incontinence.

Lower Extremities:
- Loss of power, change in sensation.
- Tremors; changes as above.
- Gait, tendency to fall and how.

Neurological Examination.
- General condition and appearance, attitude of head, mental condition in detail.
- Questions and answers.
- Memory.
- Speech.
- Writing (voluntary, by dictation, and copying).
- Head.

Cranial Nerves:
1. Smell (both sides).
2. Vision and color fields, scotoma, ophthalmoscopic record. Reaction to L. and A. Nystagmus.

3, 4, 6. Ocular movements.

5. *Sensory.*—Sensation of face and cornea.

5. *Motor.*—Mastication. Taste.

7. Atrophy or paralysis.

8. *Cochlear.*—Bone and air conduction.

8. *Vestibular.*—Rotation tests, caloric tests, pointing tests.

9. Taste.

10. Pulse, respiration, larynx.

11. Sternomastoid and trapezius.

12. Movements of tongue.

TESTS OF THE VESTIBULAR APPARATUS

	Spontaneous	
	Right	Left
Nystagmus Looking to right	Shoulder from above Shoulder from below Shoulder from side	
Looking to left		
Douche right Amp. After Minutes Seconds	*Caloric* To Douche right Shoulder from above	To
Douche left Amp. After Minutes Seconds	Douche left Shoulder from above	
To right Amp. Duration Seconds	*Turning* To right To Shoulder from above	To
To left Amp. Duration Seconds	To left Shoulder from above	

Falling reactions.
  Spontaneous.
  After stimulation.

Subjective phenomena
- Vertigo
  1. Spontaneous.
  2. After stimulation.
  3. Systematized.
- Nausea

Upper Extremities:
  Power, movements, ataxia, tremors.
  Sensations of pain, touch, temperature.
  Deep sensations and joint sensations.
  Muscular atrophy. Reflexes, astereognosis, apraxia, atopognosis, adiadokokinesis.
Abdomen:
  Appearance.
  Reflexes.
  Sensations.
  Cremaster reflex.
Lower Extremities:
  Appearance, power, walking.
  Sensations as above.
  Reflexes, knee-jerk, ankle-jerk, clonus, toe reflexes.
Frontal:
  Mental condition.

Parietal:
  Convulsions, paralysis, astereognosis, apraxia, subjective sensations.
Occipital:
  Subjective sensations of vision.
Temporal:
Hippocampal:
  Subjective sensations of smell, taste, hearing, dreamy state.
Cerebellar:
  Ataxia, adiodokokinesis, tendency to fall in any particular direction.
Hypophysis:
  Signs of hypo- or hyperpituitarism.

Sensory Examination (Head and Holmes)

(*A*) Spontaneous sensations.
    Pain, numbness, tingling.
    Pain of the limb. Idea of the limb.
(*B*) Loss of sensation.
    1. Touch.
        (*a*) Light touch.
            Cotton-wool on hairless and hair-clad parts.
            Threshold with von Frey's hairs.
        (*b*) Pressure touch.
            Threshold with the pressure esthesiometer.
    2. Localization:
        Naming the part touched.
        Henri's method as modified by Head and Holmes.
    3. Roughness:
        Threshold with Graham-Brown's esthesiometer; sandpaper tests discrimination of relative roughness.
    4. Tickling and scraping.
        Tickling on soles and palms.
        Cotton-wool, rubber over hair-clad parts.
        Light scraping with the finger nails.
    5. Vibration:
        Loss of diminution of sensibility.
        Alteration in the character of the sensation evoked.
    6. Compasses:
        Points simultaneously applied.
        Points successively applied.
    7. Pain:
        (*a*) Superficial pain.
            Pin-prick.
            Threshold with the algesimeter.
            Reaction to measured painful stimuli.
        (*b*) Pressure pains.
            Threshold with the algometer.
            Reaction to painful pressure.

8. Temperature:
    - Threshold for heat and cold.
    - Effects for adaptation on the threshold.
    - Discrimination of different degrees of heat and cold.
    - Affective reactions: (*a*) To extreme degrees; (*b*) to warmth.
9. Positions:
    - By imitating with the sound limb the position of the affected limb.
    - By pointing with the sound limb.
    - Measurement of defect by Horsley's method.
10. Passive movement:
    - Appreciation of movement.
    - Recognition of direction of movement.
    - Measurement of the angle of the smallest movement which can be appreciated.
    - Falling away of the unsupported limb when the eyes are closed.
11. Active Movement:
    - Imitation of movement by sound limb.
    - Ability to touch known spot.
    - Measurement of the defect by Horsley's method.
12. Weight:
    - (*a*) With the hand supported:
        - Recognition of difference of weight applied successively to one hand.
        - Appreciation of increase or decrease of weight.
        - Comparison of two weights placed in one hand.
    - (*b*) With the hand unsupported:
        - Comparison of two weights, one placed in each hand.
        - Recognition of difference in weight applied successively to one hand.
13. Size:
    - Difference—Threshold.
    - Distinction of the head from the point of a pin.
14. Shape (two dimensional).
15. Form (three dimensional).
    - Recognition of common objects by their form.
16. Texture.
17. Dominoes.
    - Ability to count points by touch.
18. Consistency.
19. Testicular sensibility.
    - Light pressure.
    - Painful pressure.
20. Sensibility of the glans penis.

Symptoms and Diagnosis.—All the symptoms of tumor in the intracranial cavity are produced through various effects of compression. These effects may act in one of two ways, either by irritating nerve cells or destroying them. Consequently all the symptoms might be grouped under one of these two headings. In view, however, of the importance of determining the exact location of a lesion, it is primarily necessary to classify all symptoms according to their localizing value.

For this reason we must recognize that a tumor may produce two types of symptoms:

1. General.
2. Focal.

Either type may be present in a given case or both types may be observed together. Whether the symptoms are general or focal depends to a considerable extent upon the nature of the tumor. If it is an infiltrating tumor and therefore one that replaces tissue without displacing it, focal symptoms without general symptoms are more commonly encountered, but if it is a non-infiltrating tumor it may compress the brain without replacing it, and in this case general as well as focal symptoms may occur.

The *general* symptoms are so called because they are an evidence of increased intracranial pressure but give no clue as to the location of a lesion. It is the smoke which indicates that a fire exists but does not point out where it is.

When these general symptoms exist they are of great value but it must be realized at once that *their partial or complete absence in children is one of the most important facts to be kept in mind in studying brain tumors in children.* The "general" symptoms are dependent upon the increase of intracranial pressure in a rigid closed cavity. In young children the cavity is not closed as the sutures are still open, while in older children the sutures give way fairly easily and thus the general pressure symptoms may be relieved. This separation of the sutures and the thinning of the bones that occurs in children (much more than in adults) is an attempt on the part of nature to carry out some of the features of a decompression operation. Unfortunately, however, the relief is only partial, and by the time the sutures have separated, the optic nerves have often been injured beyond repair.

There are three general symptoms that are commonly recognized but there are others of equal importance which are not so well known but still of great value.

Headache.—This may be intermittent or constant. It does not affect any particular part of the head and its location has no relation to the site of the tumor. The violence of the headaches is at times terrific and no drug affords relief. It is important to try to differentiate the character of the headache. At times the pain may be due to tenderness of the bone itself or due to involvement of the trigeminus. In very young children it will be impossible to determine the character of the pain or even that it is present.

Vomiting is often associated with headache and has been spoken of as being projectile, but vomiting is so commonly projectile that this can hardly be considered diagnostic. The vomiting of intracranial pressure, however, is not associated with nausea and is independent of the taking of food and this is the characteristic feature. It quite often is the forerunner of a violent paroxysm of headache.

Failing vision is due to choked disc. Except for the actual death of a patient, this is the most distressing of all the many symptoms—blindness with mental faculties unimpaired. Whenever any patient complains of failing vision an immediate ophthalmoscopic examination should be made. Such a custom would prevent untold misery. The cerebrospinal fluid is the first thing to be crowded out of the cranial cavity when the pressure increases and some of this is forced down the sheath of the optic nerve and causes interference with the circulation in the nerve. The various disturbances that follow produce the condition recognized with the ophthalmoscope as choked disc. There still is some discussion as to whether choked disc is due to pressure but the admirable work by Cushing and Bordley,* Wilbrand and Sänger,† leaves no room for doubt. If the choked disc is permitted to continue any length of time, atrophy of the nerve takes place and loss of vision ending in complete blindness follows. The degree of swelling in the two eyes may be different; Horsley‡ believed this to be of localizing value in determining the side on which the tumor was. Paton§ reviewing his cases, however, does not find this is the case. It is important to distinguish between the amount of swelling of the disc and the age of the process. In the development of a choked disc the edema progressively increases and the amount of swelling is determined by the height of the surface of the disc above the retina, expressed in diopters. The longer the swelling persists, the greater the histologic changes that take place. Thus the eye in which the process is more recent may show more swelling while the other eye in which the process has been longer may show more histologic change but less swelling. The eye in which the process is older may have less swelling. From the study of a large group of cases, however, it would seem that this sign is only of localizing value if the tumor is situated far forward in the cranial cavity. The farther back the tumor is situated, the less value is such a difference in the choked discs, while if the process is subtentorial it has no value at all. How marked the histologic changes become in a long standing case, with little swelling present, is best illustrated by albuminuric retinitis which is probably also due to pressure.

The other symptoms not so well known but equally valuable and noteworthy are:

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* Cushing, H. and Bordley, J., Observations on experimentally induced choked disc, Johns Hopk. Hosp. Bull., xx (1909), p. 95.

† Wilbrand u. Sänger, "Die Neurologie des anges." Wiesbaden (1900).

‡ Horsley, V., The operative treatment of choked disc, The Ophthalmoscope (September, 1908).

§ Paton, Leslie, A clinical study of optic neuritis in its relationship to intracranial tumors, Brain, xxxii, p. 65.

Sighing Respirations, Yawning, and Picking at Bed Clothes.—These are extremely common and quite uncontrollable on the part of the patient. Even if the patient has had a long restful sleep he may yawn continually. These symptoms are seen where the pressure is very great and usually indicate medullary compression. Picking at the bed clothes or picking the nose are symptoms for which no explanation has been offered, yet they occur with great frequency.

Mental dullness is also commonly an accompaniment of increased pressure. The patient may be difficult to arouse and give confused answers, his memory may be poor and he may forget recent events. At times he will not carry out orders though he understands what is said to him; this sometimes may simulate a confusional state and may lead the physician to think he is dealing with a mental case. It is true that sometimes these marked mental symptoms are an evidence of frontal lobe disturbance, but far more frequently they are merely a general symptom of pressure and of no localizing value. Associated with this mental dullness and lack of attention, there frequently occurs urinary incontinence. This may be so troublesome that in debilitated individuals it becomes a grave nursing problem.

Paralysis of the Sixth Nerve.—One of the most common symptoms is paresis or complete paralysis of *one sixth* nerve. For many years neurologists have attempted to make use of this symptom in localization, but careful anatomical studies by Cushing* have developed certain anomalies about the course of the sixth nerve in the cranial cavity; these anomalies have not only explained its frequent involvement but have also established that it is a general symptom of pressure and not a focal one. The sixth nerve has the longest course of any nerve in the cranial cavity and therefore is more likely to be subjected to pull or distortion. In addition, as it comes out of the pons, at times it is pressed upon by one of the branches coming off the basilar artery. This pressure is great enough to cause a paresis of the nerve and if it continues long enough may cause a complete paralysis. As the intracranial pressure fluctuates the degree of paresis may vary. It is important to remember, however, that paralysis of *both sixth nerves* is not a general symptom but a focal one. When one considers the frequency of brain tumors and the frequency of anatomical variations, it is highly improbable that a case of brain tumor would have the anatomical variation on both sides. It is far more likely to be due to a lesion at some point where the sixth nerves are close together, namely, in the pons, at their origin. I have seen three cases of bilateral sixth nerve paralysis and in each the tumor involved both sixth nerve nuclei.

In young children all these general symptoms may be absent due to the giving way of the sutures.

Focal Symptoms.—The character of a focal symptom depends upon the region of the brain which is affected. These symptoms may be as varied and as numerous as the centers in the brain. Any center that is affected may give rise to a focal symptom. If the tumor merely irritates it without destroying it, the symptom will partake of the function of that center but be abnormal in character, for instance a patient with his taste center affected constantly thought he tasted blood when there was nothing in his mouth; but if the tumor destroys the center, that particular function will be lost. There still remain a number of areas in the brain whose function is not known and these are spoken of as *silent areas;* consequently it may happen that a tumor may start in a silent area and reach considerable size before it produces any symptoms. Therefore a focal symptom does not necessarily mean that the tumor has begun in that region but that that is the first symptom-producing-area which has become involved.

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* Cushing, H., Strangulation of the nervi abducentes by lateral branches of the basilar artery in cases of brain tumor, Brain, xxxiii (1910–1911), p. 204.

Frontal Lobe.—Tumors in this region may produce a motor speech disturbance. In right-handed individuals the motor speech center is located on the left side while in left-handed individuals it is on the right side. There is some question whether the speech center is unilaterally located in young children. Mental disturbances occur, changes in disposition, and personality. These changes may be so subtile that they are not remarked for a long time. Several cases of kleptomania have been reported and this tendency to steal developed synchronously with the tumor. Lying, noisiness, unreasoning hilarity (witzelsucht) have all been noted.

Every institution for the insane contains a number of cases of frontal lobe tumors that have been sent there because of their mental status. Frequently the diagnosis is made even more difficult through the absence of all general symptoms. Eye ground changes are often absent because a frontal lobe process does not interfere with the cerebrospinal circulation and therefore there is no choked disc but instead there may be a primary optic atrophy due to direct pressure on the optic nerves. In rare cases conjugate deviation of the eyes may be produced by an irritative lesion over a center in the second frontal convolution which controls these movements, described by Beevor and Horsley.

If the process lies far back in the frontal lobe on the upper surface near the longitudinal sinus the abdominal reflex may be lost (Stewart and Holmes) on the opposite side of the body without any other evidence of involvement of the precentral area due to the fact that the center for the abdominal muscles lies farthest forward.

Parietal Lobe.—There are three groups of functions in this region that may give rise to symptoms. The motor area lying in the precentral gyrus, the post-central gyrus which contains the cortical centers for general sensation, and the area behind this controlling the stereognostic sense (the ability to recognize objects by touch) (Mills).

If the lesion does not immediately destroy an area in the precentral gyrus but only irritates it, focal epileptic convulsions (Jacksonian fits*) are induced and the aura is a motor one. Frequently only the aura occurs without a general convulsion following it. At other times, if the cortical irritation is severe enough, the aura is followed by a spreading convulsion and at times by a period of unconsciousness. If the primary lesion is over the post-central gyrus the aura will be sensory, but on account of the close proximity of the motor centers, convulsive movements follow if the irritation is strong enough to spread to neighboring centers.

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* Jackson, Hughlings, On the scientific and empirical investigation of epilepsies, Med. Press and Circular, xviii (1874); xix; xx; xxi; xxii (1875).

A Jacksonian convulsion spreads in a typical way and involves successive centers. Thus if the aura is in the thumb the convulsive movements involve successively the fingers, forearm, arm, and then go either up to the face or down to the lower extremity. At times a tumor first produces a paralysis and Jacksonian convulsions only appear later. This is particularly the case if tumors develop in the cortex or subcortical region. Dural tumors pressing on the cortex, on the other hand, usually produce convulsions first and the paralysis only develops when the tumor has attained considerable size. The sequence of paralysis and convulsions constitutes a valuable differential point in diagnosis. Paralysis first, followed by convulsions, points to the tumor being cortical or subcortical, while the opposite sequence strongly suggests a dural tumor.

The proper evaluation of sensory disturbances due to cortical disease has been one of the most difficult problems confronting clinicians. It of course can only be studied on human beings; animal experimentation is of little value though attempts have been made by Dusser de Barenne* and others. The most accurate information has been obtained recently from the study of a group of wounded soldiers. According to these studies "the sensory activity of the cortex may be divided into three groups. The cortex endows sensation with three discriminative faculties, (*a*) recognition of spacial relations, (*b*) a graduated response to stimuli of different intensity, (*c*) appreciation of similarity and differences in external objects brought into contact with the surface of the body" (Head†). Similar studies have also brought out the symptoms arising from the subcortical sensory pathways down to the optic thalamus (Head, Sachs,‡ Roussy§).

The accurate sensory examinations required to determine these disturbances cannot be carried out at all on young children and only with advantage on intelligent individuals. Any slowing of mental processes as commonly occurs with increased intracranial pressure may also interfere with an examination of this sort. Where the tests can be applied they make a more accurate localizing diagnosis possible. Such an accurate localization will be of value to the surgeon in deciding upon the location of the field of operation as well as in the matter of prognosis. Naturally the more accessible the tumor the better the prospect of removal.

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* Dusser de Barenne, J. G., Experimental researches on sensory localizations in the cerebral cortex, Quart. Journ. of Exp. Physiol., ix (1915–1916), p. 355.

† Head, H., Sensation and cerebral cortex, Brain, xli, p. 57.

‡ Sachs, E., The structure and functional relations of the optic thalamus, Brain cxxvi (1909).

§ Roussy, G., "La Couche Optique," Paris (1907).

The third function of this region is that of stereognosis. The center controlling this function lies between the upper portion of the post-central gyrus and the parieto-occipital fissure.

The occipital lobe contains the cortical center of vision. This center is located both on the external surface as well as the mesial surface of the brain. Irritation of the cortex produces subjective visual disturbances. These are usually in the nature of colored or white flashes of light which appear to arise from the side of the lesion, which at times are so vivid that the patient can describe them with great accuracy. At other times the visual sensations take the form of figures of persons or objects. These sensations constitute the visual auræ of Jacksonian convulsions and in every case the aura is

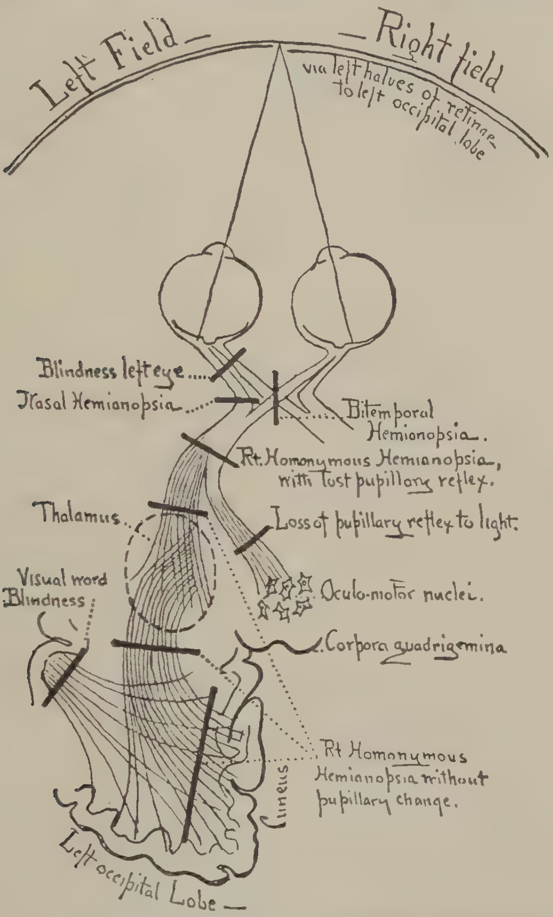
Left Field —
— Right field
via left halves of retinae to left occipital lobe
Blindness left eye
Nasal Hemianopsia
Bitemporal Hemianopsia.
Rt. Homonymous Hemianopsia, with lost pupillary reflex.
Thalamus
Loss of pupillary reflex to light.
Visual word Blindness
Oculo-motor nuclei.
Corpora quadrigemina
Rt Homonymous Hemianopsia without pupillary change.
Cuneus
Left occipital Lobe —

FIG. 16.—Visual pathway showing types of disturbances in various parts of the tract. (Cushing.)

always the same. Whenever a destructive lesion of the occipital lobe occurs, various types of homonymous hemianopsia develop, for the same half of each retina is represented in one cortex. Thus the right occipital lobe supplies the right side of each retina and the left lobe the left. There may be lateral, superior, or inferior quadrantal defects, according to the portion of the cortex which is affected. Whether the macula field had unilateral or bilateral representation was long a disputed point but it seems quite certain now that its representation in both cortices is similar to that of the rest of the visual field (Holmes*).

Distinguishing lesions (Fig. 16) of the visual cortex from those in the optic radiation or in the visual pathway more anteriorly is most important. The following points are of value: First, subjective visual disturbances only arise in the cortex; second, lesions in the occipital lobe present the Wernicke pupillary phenomenon, that is, the reaction of the pupil to light even when light is thrown on the blind portion of the retina; third, complete occipital lobe blindness is unattended by any pupillary disturbances; fourth, lesions far forward in the optic radiation, if they involve the region known as Wernicke's field, have subjective sensations of smell or taste associated with them; this is due to the fact that the fibers from the temporal and hippocampal lobes all come together in that region; fifth, lesions far forward in the optic radiation, especially if the process comes near the surface, may produce word deafness.

The temporal lobe. Much of the temporal lobe is a silent area of the brain and tumors may become very large before they give rise to symptoms. There may, however, occur auditory aphasia in right-handed individuals if the process is on the left side. There may also be subjective sensations of smell or taste for the cortical centers for these functions are located in the tip of the temporal and hippocampal lobes. These symptoms are observed most frequently in pituitary tumor cases when they leave the sella turcica and spread laterally and impinge on the temporal lobe. Involvement of the temporal lobe also brings on the attacks spoken of as "dreamy state." This peculiar series of symptoms is characterized by a sense of unreality with loss or change of personality (Kennedy†). At times patients with temporal lobe tumors are strangely inert, difficult to arouse, and unresponsive. This may be merely a more aggravated form of the dreamy state, but is so like the general symptoms associated with increased intracranial pressure that it is difficult to distinguish. A still further symptom that is only discovered after most careful examination is the loss of a small group of fibers which supply the inner lower portion of each retina. These run a very circuitous course through the temporal lobe as pointed out by Adolph Meyer. Tumors in the temporal lobe may give rise to auræ of smell or taste which are followed by focal motor convulsions. In such cases the convulsion always begins in the face and travels down the body picking out successive centers along the precentral gyrus.

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* Holmes, G., Cortical localization of vision, Brit. Med. Journal, ii (1919), p. 193.

† Kennedy, Foster, The symptomatology of tempero sphenoidaltumors, Arch. of Int. Med., Chicago, viii (1911), p. 317.

The Cerebellum, Pons and Medulla (Figs. 17 and 17*A*).—The symptoms arising from this region vary a great deal depending on where the process starts. If the process starts near the aqueduct of Sylvius the outflow of cerebrospinal fluid may be interfered with at an early stage and therefore general pressure symptoms constitute the most

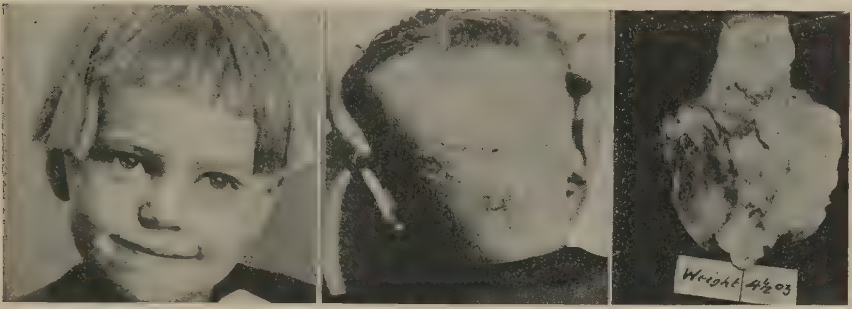

Fig. 17.—Patient from whom glioma of the cerebellum was removed in a two stage operation.

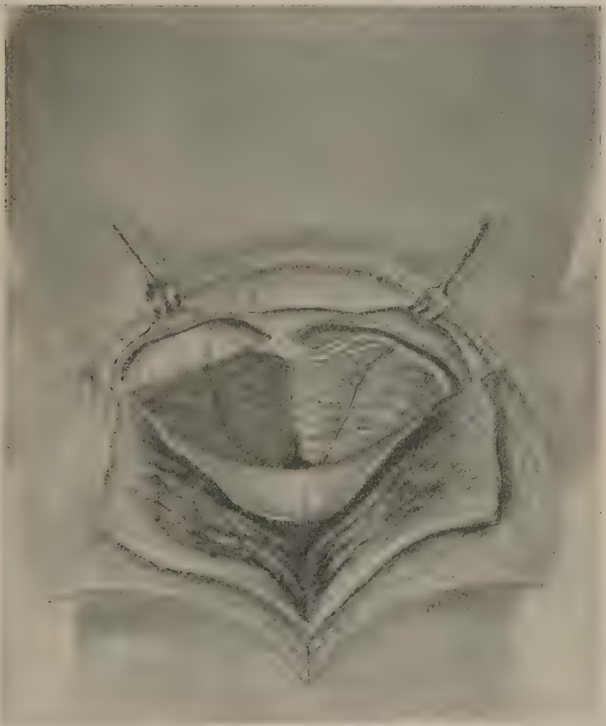

Fig. 17*A*.—Location of tumor in the cerebellum in patient shown in Fig. 17.

prominent aspect of the clinical picture and may obscure any focal symptoms. If the process, however, starts in the cerebellum, well away from the aqueduct, the symptoms may be purely focal. The cerebellum has as its principal functions the control of the finer movements of coördination, both of the upper and lower extremities. When the upper extremities are affected, there may be a tremor or ataxia or difficulty in carrying out delicate coördinated movements, as are needed for piano playing. When the lower extremities are affected, there is difficulty in walking and a staggering gait. When the patient displays this cerebellar gait, the picture is a very striking one. The legs are held far apart, he is said to walk on a broad base, and sways from side to side, at times falling down. Frequently the patient tends ts fall to one side or falls backwards or forwards. This may be of considerable localizing value. The patient falls to the side of the lesion, thus in right sided lesions he falls to the right. Falling forward or backward is an evidence that the lesion is in the vermis though the proof for this is not as strong as in the case of lesions of the lateral lobes. The mechanism of the semicircular canals is connected with the vestibular portion of the eighth nerve in the medulla and the cerebellum. The examination of this mechanism has been of great interest to the otologists in recent years and they, led by Barany* and Jones,† have devised elaborate methods of studying this mechanism. Stimulating the normal vestibular mechanism of one side produces three types of symptoms, nystagmus, vertigo and past pointing. (By past pointing is meant that an individual when blindfolded does not touch an object held before him but always deviates to one side.) In a pathologic case these symptoms may be in part absent or vary from the normal. With these facts the otologists localize a lesion in the pons, medulla or cerebellum. In children under four these tests cannot be applied as a certain amount of intelligent coöperation from the patient is necessary. In using these tests for focal diagnosis great care must be exercised (Sachs, E.‡). First of all it must be remembered that these tests after all are merely an examination of one part of the eighth cranial nerve and that therefore no diagnosis can properly be based on these findings alone. Secondly, internal hydrocephalus is a very common accompaniment of posterior fossa lesions; the pressure of the fluid on the floor of the fourth ventricle interferes with the function of the tracts of the acoustic system and gives the symptoms of a focal lesion. At present the value of these examinations is somewhat overestimated.

Pathology.—Though many varieties of tumors are found, since there is both ectodermal and mesodermal tissue in the cranial cavity, gliomata and endotheliomata greatly preponderate.

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* Barany, R., Ueber vestib. apparat, Monatschr. f. Ohrenh., xi (1906), pp. 193–297.

Barany, R., Beitr. zur Lehre von den Funktionen der Bogengänge, Zeitschr. f. Psychol. u. Physiol. d. Sinnesorgane, xli (1906), p. 371.

† Jones, I. H., "Equilibrium and vertigo." Lippincott, Phila. (1918).

‡ Sachs, E., "Annals of otology, rhinology and laryngology." (March, 1919).

Glioma is a type of tumor that only develops in nervous tissue from the supporting tissue of the brain and occasionally but very rarely in the spinal cord and peripheral nerves. This type of tumor is usually considered a malignant one and since this type is so very common it has created a very gloomy picture for the prognosis of brain tumors. Pathologists differ somewhat in their definition of malignancy, one group calls metastasis the proper criterion, the other group is guided by the invasive character of the tumor. In view of the peculiar physical properties and structure of brain tissue, infiltration cannot, in my opinion, (Sachs*) be considered a proper criterion of malignancy, and gliomata never metastasize. For this reason I believe we are justified in not calling a glioma a malignant tumor. The gliomata may be divided into three types depending on the character of the cells as suggested by Tooth.†

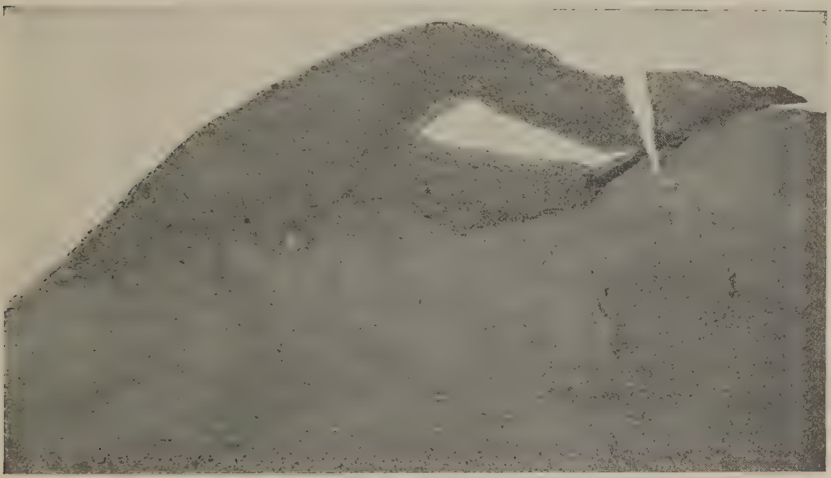

Fig. 18.—Glioma in which glia nuclei predominate.

1. Those composed of glia nuclei.
2. Those containing giant cells.
3. Those containing a preponderance of glia network and a few cells (Figs. 18–20).

These tumors are therefore to be included in the operable class though it is imperative that they be operated early and very radically.

Endotheliomata.—These tumors always arise from endothelial cells and as a rule develop from the dura but occasionally from the ependymal cells of the ventricles. The tumors are well encapsulated and grow slowly and at times attain enormous size. They are among the most favorable operative cases we have and even when very large may be removed with comparatively little difficulty. Occasionally these tumors contain calcified deposits and occasionally the skull also may be thickened over the site of the tumor; both of these conditions show

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* Sachs, E., A review of eight years' experience with brain tumors, Archives of Surgery, i, No. 1, pp. 74–84.

† Tooth, H. H., Some observations on intracranial tumors with special reference to gliomata, Brain, xxxv, p. 61.

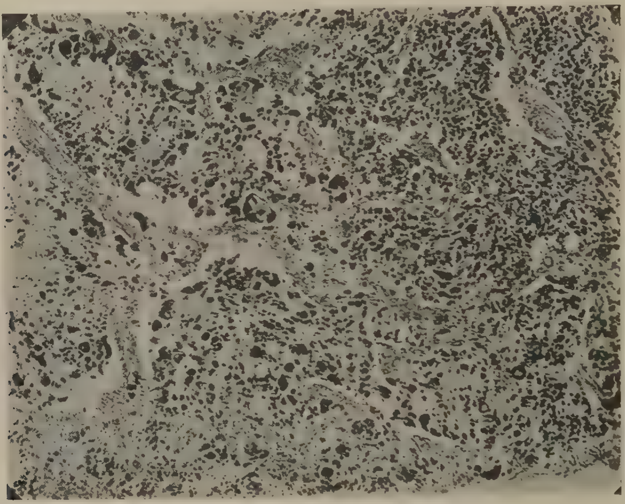

FIG. 19.—Glioma containing giant cells.

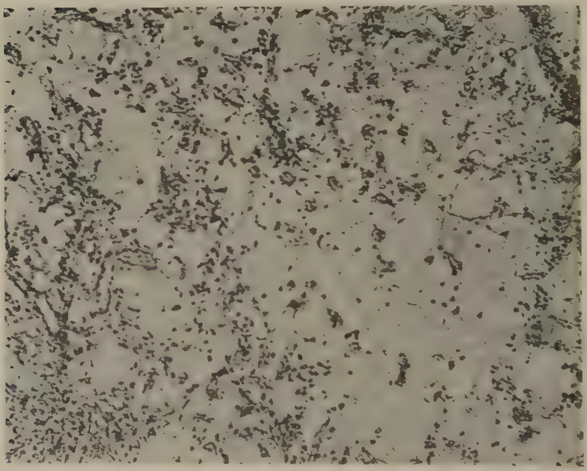

FIG. 20.—Glioma in which glia network predominates.

in an x-ray picture. Endotheliomata grow so slowly that the brain may accommodate itself and no general pressure symptoms be present. Even focal symptoms may be entirely absent and the sole clue to the condition may be the x-ray plate, as in a recent case of mine that had had general epilepsy for 15 years and no focal or general signs except headache which had developed about four weeks before admission. The tumor immediately after removal weighed 70 gm. (Fig. 21).

Other varieties of tumors may be found in the cranial cavity; of these the chief are fibroma, lipoma, sarcoma and angioma. The pathology of all these conditions presents no unusual features when they occur in the cranial cavity. In this group must be included the granulomata, particularly tuberculoma or solitary tubercles. Though tuberculosis most frequently presents the picture of tuberculous meningitis when it attacks the nervous system, in a certain percentage of cases it forms large localized tuberculous masses which are well encapsulated. Such a tuberculoma produces the picture of a true brain tumor. The major part of the mass is composed of necrotic caseous tissue with giant cells and characteristic tubercles about the margin.

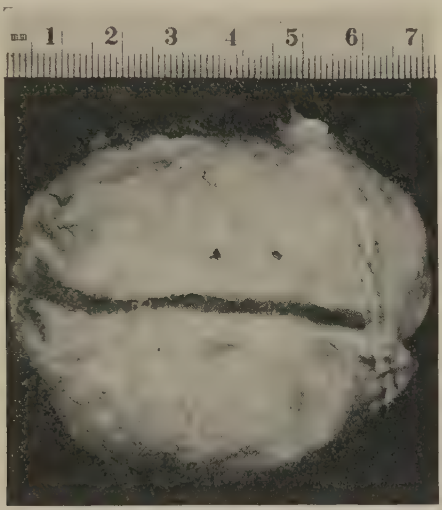

Fig. 21.—Dural endothelioma weighing 70 gm. Removed from patient who had no symptoms of intracranial pressure except headache. Tumor present about 15 years.

Metastatic processes occur in the brain as elsewhere; they as a rule are carcinoma and enter the cranical cavity through the blood stream. The areas are multiple and present a confusing clinical picture. The finding of a primary focus elsewhere establishes the diagnosis.

Pneumoventriculography.—(Ventriculography, air injection.) In 1918 Dandy[*,76] first described a new diagnostic method for localizing brain tumors. Since then, numerous other publications on this subject have appeared by Dandy and others. It consists in withdrawing cerebrospinal fluid from the ventricles and replacing it by air. The *x*-ray picture of such a case is startling. Dandy pointed out that tumors deform the ventricles and thus one may be able to determine the location of tumors otherwise unlocalizable. The method is an

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[*] Dandy, W. E., Ventriculography following the injection of air into the cerebral ventricles, Annals of Surgery (July, 1918), pp. 5–11.

important addition to our diagnostic armamentarium. In certain cases it is a great help and enables the surgeon to know exactly where the tumor is. The presence of internal hydrocephalus, the position of the ventricles in relation to the median line, and the dilatation of one ventricle or deformity of a ventricle, are the principal points that are made out in the x-ray plates. It must be emphasized, however, that the method is dangerous and has a mortality of at least 3 to 5 per cent. in the most careful hands. It should only be undertaken by those familiar with intracranial surgical procedures. Another point of great importance is that the x-ray pictures at times are very misleading, as deformations of the ventricles occur normally. I have operated several times on the strength of the findings in an air injection and not found the tumor at the site indicated. After six years experience with the method, I feel that it is at times of great value. It should only be used where neurological examination fails to localize a lesion. It is by no means infallible, and on account of its attendant danger should be used with great caution.

Pituitary Tumors.—Though these tumors are essentially brain tumors their symptoms and clinical course differ so widely from those of brain tumors in general that they must be considered independently.

Anatomy and Physiology.—The pituitary gland lies in the sella turcica covered by a layer of dura and surrounded on each side by the cavernous sinus. In front of it lies the optic chiasm and the optic tracts pass back on either side of the sella. The pituitary gland consists of three parts, the anterior lobe, posterior lobe, and pars intermedia. Springing out of the gland in the region of the pars intermedia is the stalk which opens into the third ventricle and through it, according to Cushing,* products of the pituitary gland are discharged into the cerebrospinal fluid. The anterior lobe is concerned principally with bony growth while the posterior lobe is concerned with sugar metabolism, and contains substances that raise blood pressure, cause diuresis and contraction of smooth muscle, notably the intestine and uterus. The gland normally undergoes hypertrophy during pregnancy and undergoes retrogressive changes which are best seen in hibernating animals.

Each portion of the gland under certain conditions may hypo or hyperfunctionate. Knowing the functions of the gland as described above, it is no difficult task to determine the pathologic physiology of the gland.

A patient may have:

1. Hyperfunction of the anterior lobe.
    Increased bony growth.
2. Hypofunction of the anterior lobe.
    Delayed bony development.
3. Hyperfunction of the posterior lobe.
    Polyuria.
    Loss of weight.

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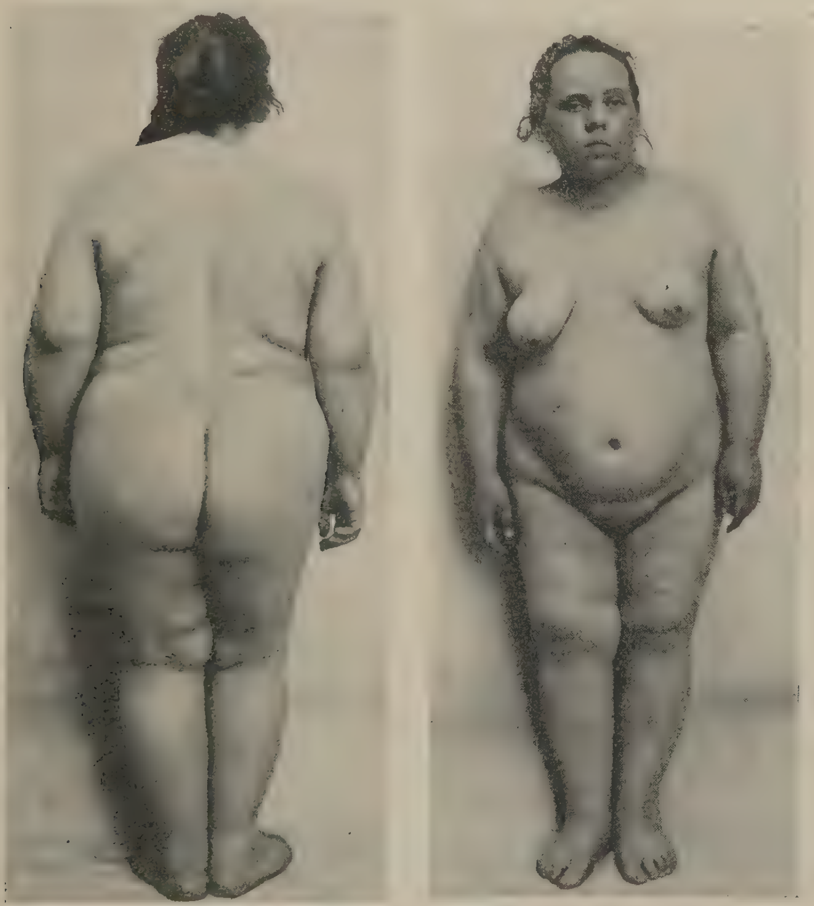
* Cushing, H., "The pituitary body and its disorders." Lippincott (1912).

4. Hypofunction of the posterior lobe.

Accumulation of fat.

Ability to store large quantities of sugar.

In addition to these symptoms, patients show a number of other symptoms which are definitely referable to disturbances of the pituitary gland but we do not know which portion of the gland is responsible for them. Some of the principal ones are:



Figs. 22 and 23.—Dyspituitarism in a girl of 14. Condition was due to bony overgrowth of the sella which compressed the pituitary.

Disturbances in sexual development.

Atrophy of the testicles, absence of the secondary sexual characteristics as hair on the body, under the arms and pubes.

Infantile appearance.

Drowsiness, at times almost approaching a condition of semiconsciousness.

Subnormal temperature.

It is a very common experience that patients present evidences of hypo- and hyperfunction of the gland at one and the same time so that one rarely sees a case which is a pure case of hyper or hypopituitarism. This is probably explained in this way. If, for example, the anterior lobe hypertrophies and hyperfunctionates, it presses on the posterior lobe and inhibits its function. Because of this mixture of symptoms Cushing suggested the term Dyspituitarism (Figs. 22, 23).

As the gland enlarges, either due to a simple hyperplasia or as a result of a new growth, it may escape the confines of the sella turcica and press on surrounding structures. These are especially the optic chiasm, the sixth and third nerves and the mesial surface of the temporal lobes, the uncinate gyrus and the hippocampal lobe. The fibers supplying the inner halves of both retinæ, therefore the ones with which we see the outer fields, lie nearest the pituitary and these are the first to be involved. This leads to that peculiar visual disturbance known as bitemporal hemianopsia. The optic nerves show ophthalmoscopically a primary optic atrophy and do not go through the stage of choked disc as occurs with other types of brain tumor.

Pressure or irritation of the uncinate gyrus leads to disturbances of smell; in these attacks which may be followed by a motor focal epilepsy the convulsion is initiated by an aura of smell.

Cushing has suggested that all cases may be divided into these four groups:

1. Cases with glandular symptoms and very few if any neighborhood symptoms.
2. Cases with neighborhood symptoms and few if any glandular symptoms.
3. Cases with marked glandular and marked neighborhood symptoms.
4. Cases with pituitary symptoms and also some referable to other ductless glands.

There are certain clinical types of pituitary disorders that occur particularly in children, though all the disturbances may occur.

Fröhlich's Disease or Adiposis-genitalis-cerebralis.*—In this condition the patient retains his infantile appearance. The sexual organs are not developed, the breasts in girls are infantile, there is no hair or a very scanty growth on their body after puberty; the menses are delayed; the patients are fat and have an abnormal sugar tolerance; they may have headaches and some neighborhood symptoms; they have thin tapering fingers. *X*-ray examination shows that there is delay in the union of the epiphysis with the diaphysis. Young persons of 18 may have no union whatever of the epiphyses of the small bones of the fingers. These cases also may show an abnormal sella turcica, larger, deeper or longer anteroposteriorly. As a rule, however, this type of case is due to a process, often a congenital one, lying above the sella, a so-called suprasellar tumor. These tumors may contain calcified particles or large colloid cysts which show on the x-ray plate.

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* Frankl Horchwart, L. v., Die Funktion der Hypophyse, Berl. Klin. Wochenschr., xlvii (1909), pp. 613, 670, 719.

Gigantism.—These patients are usually abnormally tall, at times reaching extraordinary heights, seven, eight and occasionally nine feet. They are the giants that are commonly seen in the circus. There are two types, those that have perfectly proportioned bodies but are extremely tall, the Lorain type, and others that have abnormally long arms and legs for the size of their body. In these latter cases there may be other signs of disturbed pituitary function, some of them may show evidences of glandular insufficiency and others of glandular hyperfunction. There may be neighborhood symptoms in these cases also. These cases of gigantism show symptoms of still another condition—*acromegaly.** In this condition the bony changes are confined to the bones of the face. There are a great many cases which show all stages of variation between the pure gigantism and the pure acromegaly depending on what the condition of the bones is when the disease begins. The sella turcica is greatly enlarged in these conditions and the diagnosis is readily made with the x-ray.

The condition in which hypopituitary function is the outstanding feature of the case is best seen in children with hydrocephalus. These patients are abnormally fat and under-developed and at autopsy the pituitary gland is greatly flattened out so that it could not possibly functionate normally.

All these types of pituitary disease occur in children and adolescents with the exception of acromegaly which really is a disease of adult life.

Treatment.—In order to institute intelligent treatment each case must be analyzed with great care.

If only glandular insufficiency is present, the only treatment necessary may be glandular therapy but in such a case an attempt must be made to determine the character of the lesion that is producing the symptoms. If x-ray studies show a suprasellar lesion or an enlarged pituitary fossa, surgical intervention may be deemed advisable.

If there are severe headaches, which may occur with any type of pituitary disease, only relief of pressure may be indicated, or if there are neighborhood symptoms in addition to general pressure symptoms it may be necessary to remove all or part of the tumor.

There are various operations which accomplish these ends. The pituitary gland may be approached in three different ways.

1. Through the nose, opening the sphenoidal sinus and then opening the floor of the sella turcica. There are various methods, modifications of one another, which accomplish this, notably those by Schlöffer,† von Eiselberg,‡ and Cushing. All of these methods have this in common that they approach the pituitary through the floor of the sella.

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* Marie, Pierre, "L'Acromègalie" Iconographie de la Salpetrière, i (1888), pp. 173, 229; ii (1889), pp. 45, 96, 139.

† Schlöffer, H., Zur Frage der Operationen an der Hypophyse, Beitr. z. Klin. Chirurg., Hft. 1 (1906), p. 767.

‡ Eiselberg, F. v., Operations upon the hypophysis, Annals of Surgery, lii (1910), pp. 1–14.

2. The subfrontal route first suggested by Krause* and developed by Frazier† and Cushing.

3. The lateral approach along the greater wing of the sphenoid practiced by Heuer,‡ Adson, and the writer.

4. The lateral approach under the temporal lobe suggested and carried out a number of times successfully by Horsley. This latter method is so difficult and gives such an unsatisfactory exposure that it has not been employed by others.

When the tumor has grown up into the cranial cavity it produces the general symptoms of intracranial pressure and these may have to be relieved by a subtemporal decompression operation.

Epilepsy.—No lengthy discussion of the causes of epilepsy is here contemplated since the subject has been exhaustively treated on p. 250.

In order, however, to make the surgical aspect of the subject clear, some facts have to be repeated that have been mentioned in that section.

This term is usually taken to be descriptive of a disease, whereas in reality it is a symptom that is observed in a variety of conditions. Convulsions take many different forms; they can be grouped under two headings, general and focal.

The essential differences between these two groups are that a general convulsion does not begin in any one part of the body but seems to involve the entire body almost instantaneously and the attack gives no definite warning or aura. The convulsions are not always accompanied by unconsciousness but may be characterized by very mild seizures known as *petit mal* attacks while the seizures in which the patient becomes unconscious are known as *grand mal* attacks.

The focal attacks, Jacksonian epilepsy, always begin in a particular part of the body, and have a definite premonitory sign—an aura. These attacks always begin in exactly the same way. The convulsive seizure may spread over the entire body and become then a general convulsion but the inception of the attacks always remains the same and the aura always initiates the attack.

In spite of the vast amount of study that has been devoted to this subject our knowledge remains pitifully small. I think there is no doubt, however, that the cortical cerebral centers are an essential part in the starting of an attack. A convulsion may be compared to a container in which the pressure has become too great and finally explodes. The more violent the explosion is, the more areas in the neighborhood may become involved. The differentiation of a focal from a general epilepsy is oftentimes not easy for both the patient himself and relatives may lead the physician to believe that the attacks begin uniformly in a characteristic way when the attack is really a general one.

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* Krause, F., "Chirurgie des Gehirns und Rückenmark," 2 vol. Berlin and Wien (1911).

† Frazier, C. H., Choice of method in operations upon pituitary body, Surg., Gyn., and Obst. (July, 1919).

‡ Heuer, George J., Surgical experiences with an intracranial approach to chiasmal lesions, Arch. of Surgery, i, No. 2, p. 368.

These auras may be sensory or motor in character. The sensory auras may originate in any one of the cortical sensory centers.

There may be a visual aura arising from the cortical center in the occipital lobe. Such an aura may consist of flashes of light, white or colored. These flashes may appear on one side, or the aura may consist of a peculiar sight, thus I have seen a patient whose attack was always initiated by a visual aura of a headless statue. There may be an auditory aura arising from the auditory center in the temporal lobe. The patient may hear peculiar sounds or voices, music, terrifying noises or what not. An aura of smell or taste may be present arising from one of the centers in the tip of the temporal lobe and an aura of disturbed sensation which is variously described by the patient as numbness, tingling, or formication, may originate in the post-central gyrus. The motor aura arises from one of the motor centers in the precentral gyrus. The focus originating the aura may occupy a very small region, thus it may consist of flexion of one finger or toe or twitching of one side of the face. There may be a speech aura, either sensory or motor. In the former type the patient may not understand what is said to him while in a motor speech aura he may not be able to speak or he may constantly repeat a certain word or phrase prior to his attack; then again there may be speech auras characterized by paraphasia or ill-defined confusion in the speech mechanism. There may be still other auras more difficult to recognize and to interpret; thus a "dreamy state" may arise from the tempero-sphenoidal lobe or psychic disturbances or those extraordinary movements described by Hughlings Jackson in cerebellar fits and observed by Victor Horsley* in stimulation experiments on monkeys and described by him as "progression movements."

Epilepsy may be caused by a great variety of conditions. Whether there really is such a thing as a "reflex" epilepsy I am unable to say postively but I have a very strong suspicion that reflex epilepsy and idiopathic epilepsy might well be used interchangeably since both are but cloaks for our ignorance. That we do not know the essential cause of epilepsy is undoubtedly so; even in the cases in which an organic lesion is present there is much to suggest that there is an underlying cause, something more fundamental than has yet been discovered that is responsible for the convulsive seizure. Epileptic convulsions may occur in any infectious diseases occurring in childhood, also in uremia, syphilis, meningitis, encephalitis, hemorrhage, brain tumors, pituitary tumors, and depressed fractures of the skull. The cortex of children, especially young children, is very much more irritable and more easily affected by whatever it is that produces a convulsion than the adult cortex.

When the neurologic surgeon is confronted with a case of epilepsy he must first determine whether he is dealing with a general or a focal epilepsy. Whenever possible, the patient's convulsions ought to be watched by a trained observer but to do this of course the patient must be in a hospital. It has been my practice for the past 10 years to send every questionable case into the hospital for study. Even then one may make an occasional mistake and call a general a focal epilepsy. The next question is to establish if possible the cause of the condition. There will always be many cases in which this is impossible.

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* Horsley, V. and Clarke, R. H., The structure and functions of the cerebellum, Brain, Part cxxi, xxxi (1908), p. 45.

Treatment.—If the case has a focal epilepsy it should always be operated upon and a thorough exploration of the diseased area carried out. If there is a removable condition in the affected center it should be excised and if no gross lesion is found the center should be carefully outlined with a faradic current and then excised as was advocated by Victor Horsley. In the case of sensory auras this may be difficult but the use of local anesthesia as advocated by Cushing, Dowman and others may make this possible.

The results of the surgical treatment of focal epilepsy justify the previous statement that all focal epilepsies should be explored but it must not be deduced from this that all focal epilepsies, even those where there is a localized removable lesion, are cured. Extensive statistics on this point are not available but every surgeon has had the bitter experience of operating on such cases and having the convulsions continue. Not infrequently the attacks stop for a time and then recur, in other cases they are diminished both in frequency and severity for long periods. There are several factors that have contributed to these disappointing results. The more convulsions a patient has, the more irritable does his cortex become. It is said to become epileptogenous, that is, develop the epileptic habit, so that even if a primary focus is removed the rest of the cortex remains supersensitive or hyperirritable and convulsions may continue. This hyperirritability is readily demonstrated at operation on an epileptic by the ease with which a convulsion may be started with a faradic current so weak that it could not stimulate a normal cortex sufficiently to produce the contraction of a single muscle. Another factor accounting for the poor results after operation is that the irritating focus has not been widely enough excised.

The surgical treatment of general epilepsy has been unsatisfactory and has been given up by all careful surgeons. Moreover the decompression operations recommended by Kocher have also not proven satisfactory. Though an occasional case may seem to be influenced favorably, there is no sound basis for the procedure and it should therefore be given up.

The subject cannot be left without a word about the relation of the pituitary to epilepsy. In recent years there has been a growing tendency to ascribe epilepsy to disturbed pituitary function. McKennan studied the sella turcica in a large number of epileptics and claimed the roof of the sella was frequently closed and that this might be a factor in causing epilepsy through interference with the function of the pituitary. Schwab and I studied a large number of epileptic skulls with stereoscopic plates and came to the conclusion that these observations were faulty. The appearance of a closed sella was due to the way the plate had been taken. Thus far I feel that there is no good evidence to justify the conclusion that the pituitary produces or influences epileptic seizures and certainly nothing as yet justifies any surgical measures on the pituitary to relieve this disorder.

Operations on the Brain.—It would be out of place in a treatise on neurologic surgery, in a system designed primarily for internists and pediatrists, to dwell at length on the technical details of cranial operations, the preparation of the field, the instruments used and the methods of opening the head. All this will therefore be dispensed with. Such information is better obtained in textbooks on surgery. It seems more desirable to acquaint the reader with the various types of operations, when they are indicated and how serious they are.

Palliative Operations.—Up to the present time and probably for many decades to come, a certain number of cases of brain tumor will come so late to the surgeon that the most he can hope to do is afford the patient relief from headache and prevent their going blind due to an advancing choked disc. For these cases, if the lesion is believed to be cerebral, not cerebellar, a subtemporal decompression operation is carried out. This consists in splitting the temporal muscle in the course of its fibers, opening the bone widely, that is, in a child making a circular opening with a diameter of at least 2 in., and splitting the dura in star shaped fashion so that the brain can herniate through this opening. The *dura is always left open* in a decompression operation. The muscle is closed carefully in layers to prevent the herniation from becoming too large and disfiguring. No drainage is ever used.

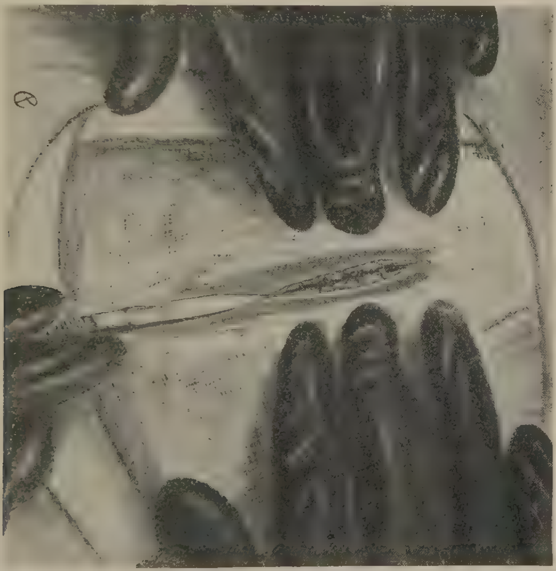

Fig. 24.—Subtemporal decompression. This shows the way in which the assistants control the hemorrhage while the incision is made down to the galea.

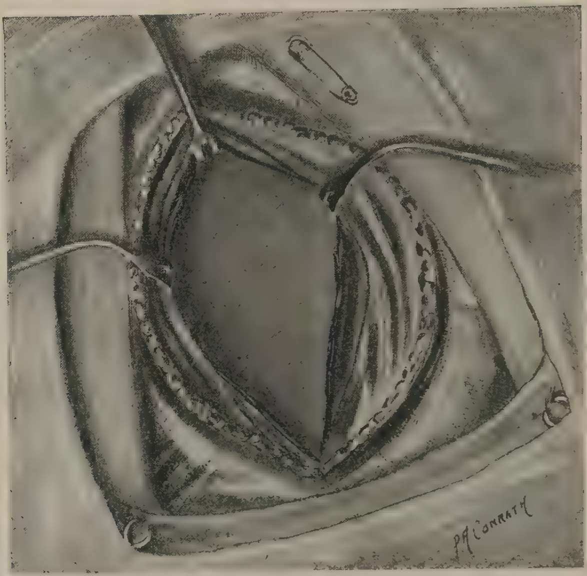

Fig. 25.—Subtemporal decompression. This shows the large area of bone that must be removed if there is much intracranial pressure in order to completely relieve the increased tension.

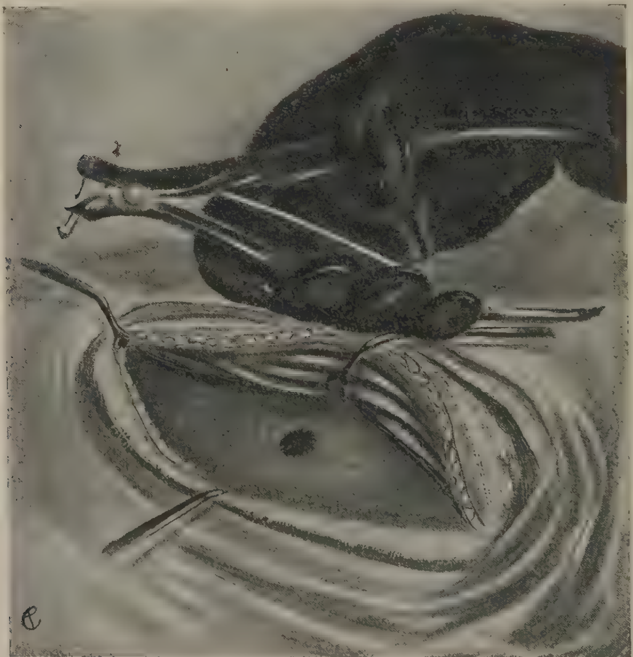

Fig. 26.—Subtemporal decompression. After a small hole has been made with a perforator and burr it is enlarged to the required size by means of rongeurs.

If the tumor is in the posterior fossa, a cerebellar decompression is done. This consists in removing bone over both lobes of the cerebellum and the posterior portion of the foramen magnum. The dura is widely opened and left open. A careful closure of the muscles follows. The usual incision is the one advocated by Cushing known as the cross-bow incision. I have in the past few years done such a decompression through a vertical incision merely splitting the muscles and retracting them. This, in children especially, gives ample exposure for a decompression.

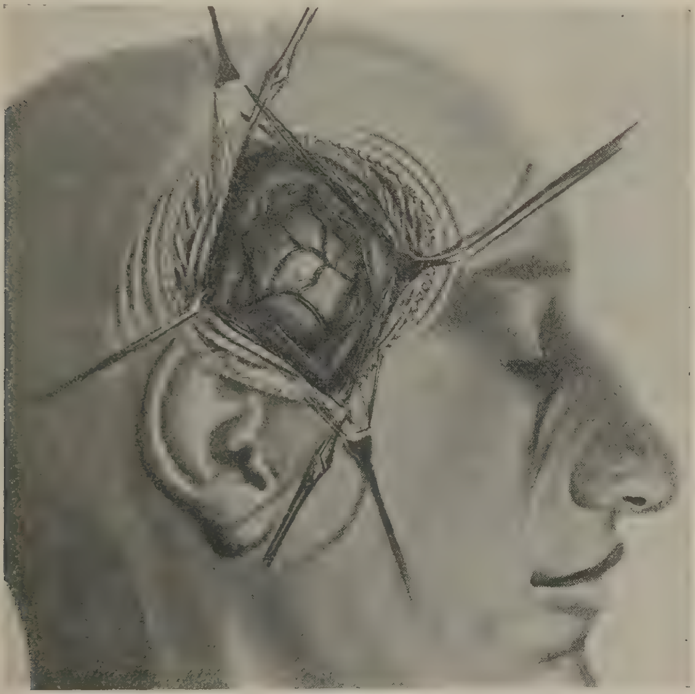

Fig. 27.—Subtemporal decompression. Showing the radiating incisions into the dura and the marked bulging of the temporal lobe. When intracranial pressure cannot be reduced prior to opening the dura, the first layer of sutures is placed as shown in the drawing before the dura is opened.

These various decompression procedures are also used in fracture cases and to relieve pressure in the case of unlocalizable tumors. The steps of the operations are shown in Figs. 24–29.

Operations for Exploration or Tumor Removal.—Whenever an area of the cerebrum is to be explored with the view of carrying out a more radical or extensive procedure, the best method is to turn down a bone flap, a so-called osteoplastic craniotomy. This is not unlike opening the lid of a box and then closing it again. After the bone flap has been reflected the dura is opened on three sides and turned back and whatever procedure on the brain deemed necessary is carried

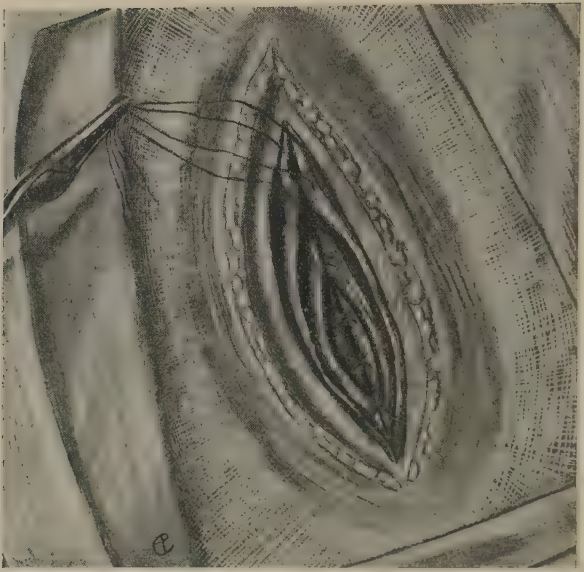

FIG. 28.—Subtemporal decompression. This shows the way the temporal muscle is sewed together in several layers prior to closing the temporal fascia. The temporal fascia must always be closed, as it prevents more effectively than the muscle, the formation of a large disfiguring hernia.

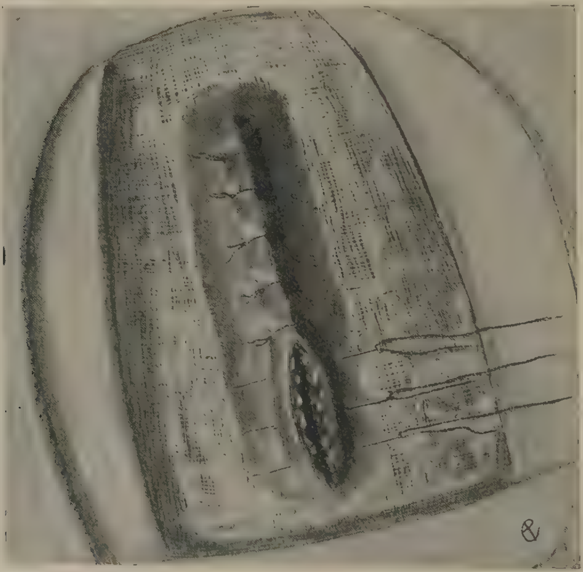

FIG. 29.—Subtemporal decompression. Interrupted sutures of fine silk are used for closing the skin.

out. The dura may be closed or if permanent relief of pressure is desirable the dura may be left open for purposes of decompression. If left open, such an opening is preferably made under the temporal muscle and the bone over it must be removed.

In the early days of cranial surgery it was *not* an uncommon procedure to discard the bone and merely stitch back the skin because the brain bulged so tremendously. This resulted in great deformity and incapacity to the patient, incapacity because the brain was merely covered by skin and galea and therefore readily liable to injury. Today every effort is made to avoid such bulging of the brain and this is best done by withdrawing cerebrospinal fluid from the ventricles.

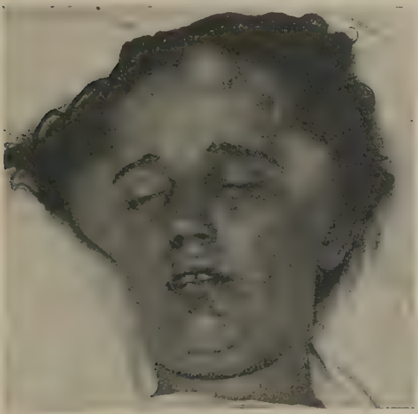

FIG. 30.

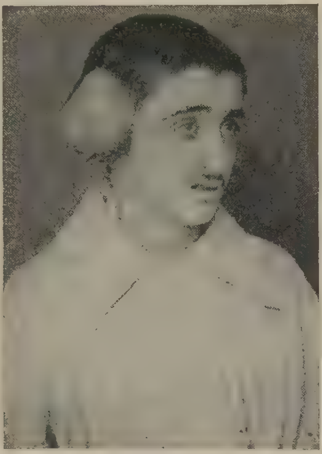

FIG. 31.

FIG. 30.—Extreme degree of herniation in a case of unlocalized tumor. There had been repeated explorations. Picture taken a few hours before death. Tumor in the septum lucidum.

FIG. 31.—Example of marked herniation following a poorly placed decompression operation. The defect in the skull had not been covered by muscle and fascia.

The question now arises, how dangerous is a decompression operation and how dangerous is an exploratory craniotomy. On this point large masses of statistics are of little value. It is far better to take the work of one clinic where uniform conditions exist. Of course it makes a great difference what the types of cases are. Tumor cases with marked pressure give less satisfactory results. The best results that have ever been obtained by anyone have been those of Cushing who reported 100 consecutive cases with a mortality of about 8 per cent. In a similar number of my own cases the mortality was about 12 per cent. These figures are quoted for but one reason, in order to emphasize the fact that cranial operations are not nearly as dangerous today as is still generally believed. These various operations are as well borne by children as by adults. There are three precautions, however, that must be insisted upon in the case of children.

First, catharsis prior to operation must be avoided; second, children very easily lose water so that it is well to fill up their tissues with subcutaneous normal salt solution before an operation; and third, to keep up the body heat. It is desirable to operate children on a heated table.

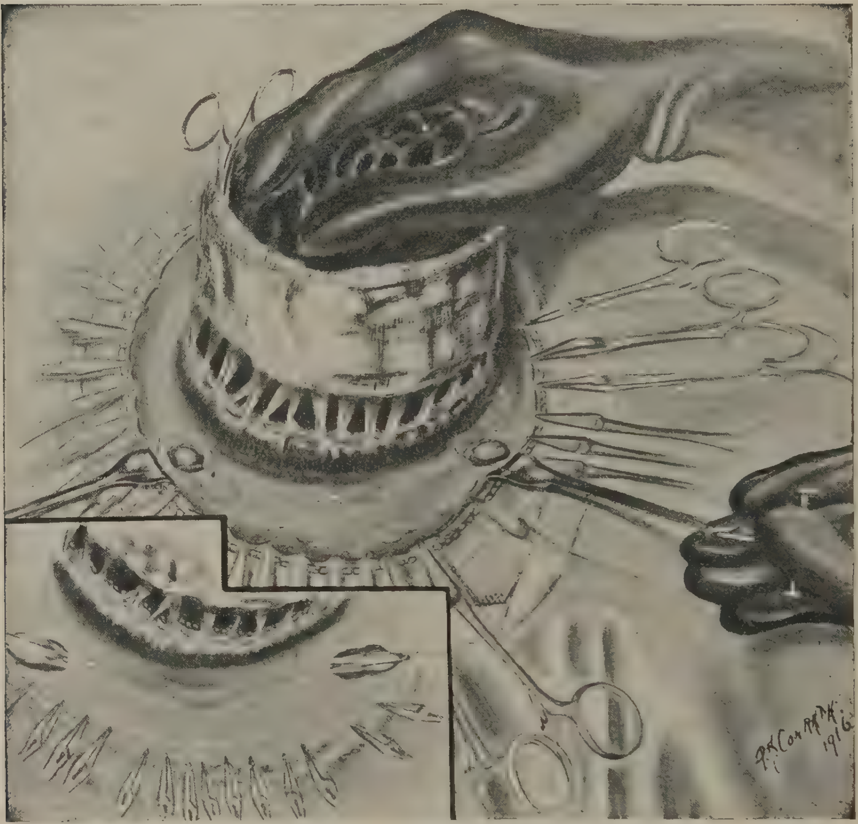

FIG. 32.—Osteoplastic flap. The edges of the bone are beveled so that the bone flap will not be depressed on the brain when replaced. The dural flap is reflected differently depending upon the location of the lesion. If no marked increased intracranial pressure is present the dura is closed with interrupted sutures (Fig. 34). If a decompression is to be combined with an osteoplastic flap, the base of the flap is rongeured away and a defect is left in the dura over the temporal lobe (Fig. 35).

Ventricle puncture is one of the most useful procedures the neurologic surgeon has at his command. The lateral ventricle may be entered with little danger from various points when the surgeon has experience. One or two precautions are necessary, first, not to injure a cortical or subcortical vessel; second, to use a needle with holes on the side so that brain tissue will not plug it; third, to pass the needle through the center of a gyrus and not through a sulcus for the ves sels all lie in the sulci. The puncture should be done where the ventricles are broadest. There are two such places, one through the frontal lobe, as advocated by Kocher, about 2 to 3 cm. to one side of the median line and a little in front of the coronal suture, or Keen's point which is at the junction of the superior and inferior cornu of the lateral ventricle about 3 cm. behind and above the external auditory meatus. There has been a tendency recently among some pediatricians to do ventricle punctures directly through the skin without opening the dura. This is an extremely dangerous procedure as one never can tell when one is going to strike a cortical vessel. Only a minute opening in the dura is necessary to convince oneself that there is no vessel in the way. Doing a ventricle puncture without incising the dura cannot be too severely condemned (Figs. 30–36).

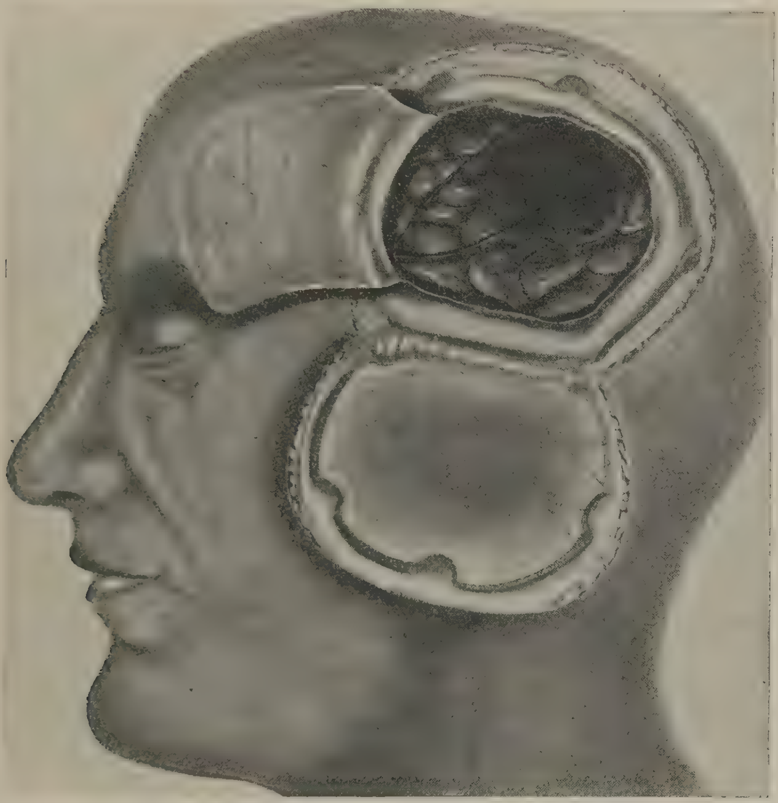

FIG. 33.—Osteoplastic flap. The edges of the bone are beveled so that the bone flap will not be depressed on the brain when replaced. The dural flap is reflected differently depending upon the location of the lesion. If no marked increased intracranial pressure is present the dura is closed with interrupted sutures (Fig. 34). If a decompression is to be combined with an osteoplastic flap, the base of the flap is rongeured away and a defect is left in the dura over the temporal lobe (Fig. 35).

NEUROSURGICAL DISEASES OF THE SPINE AND CORD

There are but a few diseases of the spine and cord in children which concern the neurologic surgeon; most of them are conditions that concern the orthopedist or general surgeon.

Congenital Anomalies.—Under the heading congenital anomalies of the brain in the section on "Brain and Meninges" I have already discussed the ones in regard to tne production of spina bifida. The condition is not a very encouraging chapter on account of the high mortality and because so many cases, even after the spina bifida is cured, develop hydrocephalus and die after a lingering illness of six months to one or two years. As we have a better understanding of hydrocephalus* than heretofore, the mere fact that a case of spina

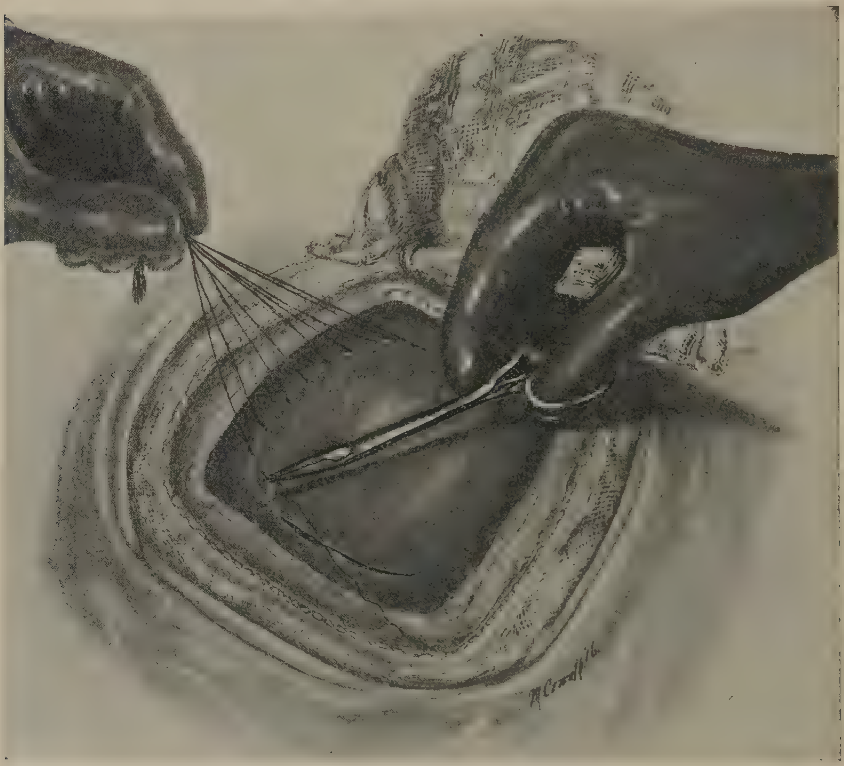

Fig. 34.—Osteoplastic flap. The edges of the bone are beveled so that the bone flap will not be depressed on the brain when replaced. The dural flap is reflected differently depending upon the location of the lesion. If no marked increased intracranial pressure is present the dura is closed with interrupted sutures (Fig. 34). If a decompression is to be combined with an osteoplastic flap, the base of the flap is rongeured away and a defect is left in the dura over the temporal lobe (Fig. 35).

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* Dandy, W. E. and Blackfan, K. D., An experimental and clinical study of hydrocephalus, Journ. A. M. A., Chicago, lxi (1913), p. 2216.

bifida develops this complication does not make it a hopeless proposition. It merely means that our efforts must be directed to cure that condition. As the type of hydrocephalus in these cases is due to hypersecretion the proper procedure would be to reduce the secreting gland, that is, removing part of the choroid plexus. In every instance it is essential to determine the type of hydrocephalus we are dealing with by the phenosulphonaphthalein test described in the section on hydrocephalus.

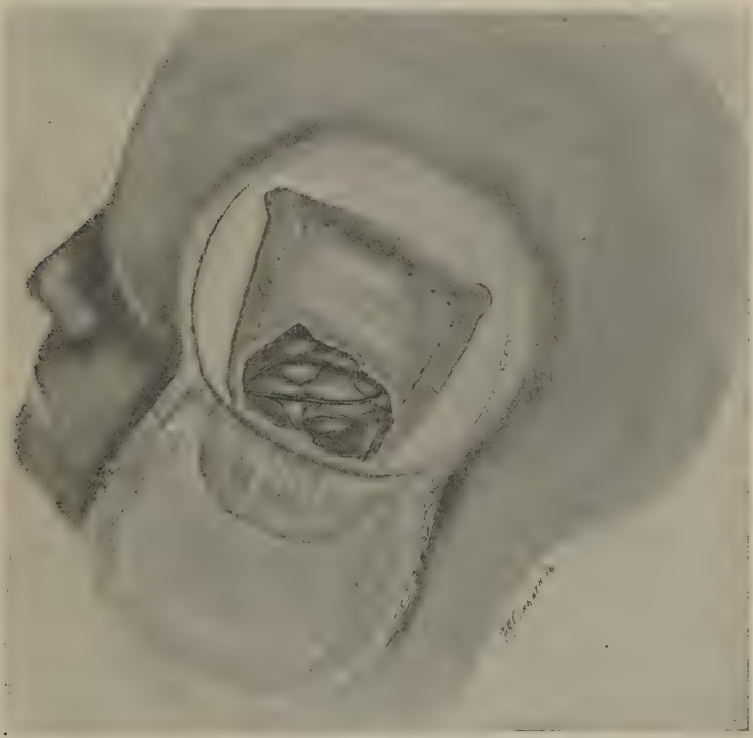

FIG. 35.—Osteoplastic flap. The edges of the bone are beveled so that the bone flap will not be depressed on the brain when replaced. The dural flap is reflected differently depending upon the location of the lesion. If no marked increased intracranial pressure is present the dura is closed with interrupted sutures (Fig. 34). If a decompression is to be combined with an osteoplastic flap, the base of the flap is rongeured away and a defect is left in the dura over the temporal lobe.

There are five types of spina bifida:

1. Rachischisis.
2. Meningocele.
3. Spina bifida occulta.
4. Myelomeningocele.
5. Syringomyelocele.

Rachischisis is that type in which the medullary canal does not close at all. It is associated with other congenital defects which make any operative procedure quite out of the question.

Meningocele (Fig. 37).—This variety of spina bifida consists primarily of a defect in the vertebral column and a herniation of the membranes of the cord. The hernia may contain only dura or arachnoid as well. The cord is developed normally in these cases as is readily determined by an examination of the extremities and the rectal sphincter. All these functionate normally. This type is the most favorable for treatment but unfortunately occurs far less frequently than the other types. The hernial sac contains cerebrospinal fluid. At times the nerve fibers have floated up into the sac and are adherent to the sac wall so that it may be difficult to determine if one is dealing with a true meningocele or a meningomyelocele. The skin over a meningocele is as a rule very thin, almost translucent. The tumors always

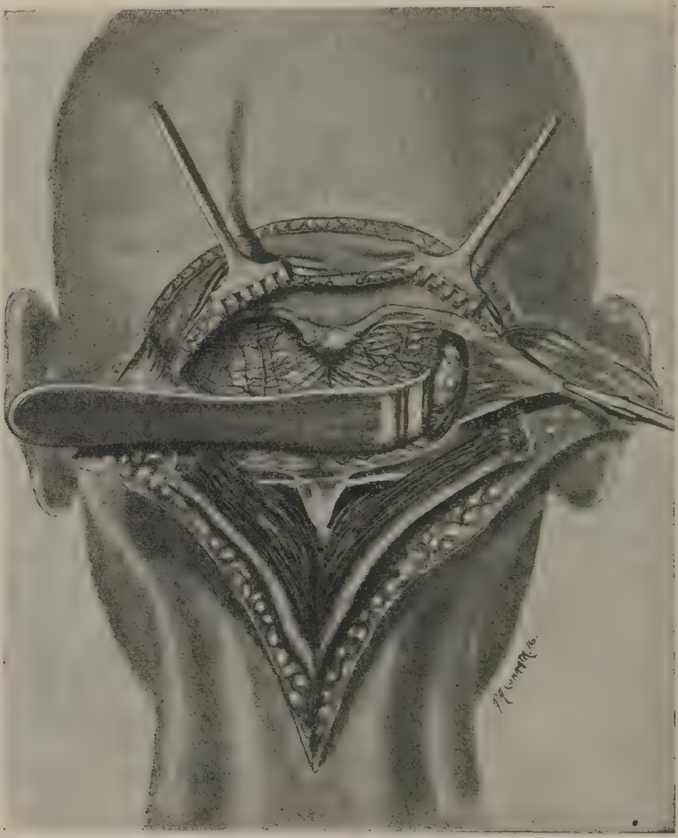

Fig. 36.—Typical cerebellar exposure showing the cross-bow incision and bilateral exposure of the cerebellum. To get a good exposure it is always necessary to expose both sides.

pulsate and fluctuate. The skin is rarely normal over the hernia, occasionally it may contain a tumor which develops from rests of the mesodermic or ectodermic layer.

Spina Bifida Occulta.—This type is as a rule a subvariety of meningocele but presents a very different clinical picture. It is often not observed until adult life. There may be no visible protrusion, only hypertrichosis at the site of the defect. In doubtful cases an x-ray plate shows a bony defect in the spine and thus the diagnosis is established. This condition is at times the cause of obscure trophic disturbances in the feet in children or young adults. The defect in the spine may contain a hernial protrusion which only contains membranes (meningocele) or nerve elements which puts it into the class of meningomyeloceles. Such a deformity may be suspected if there is a profuse growth of hair in the middle of the back (hypertrichosis).

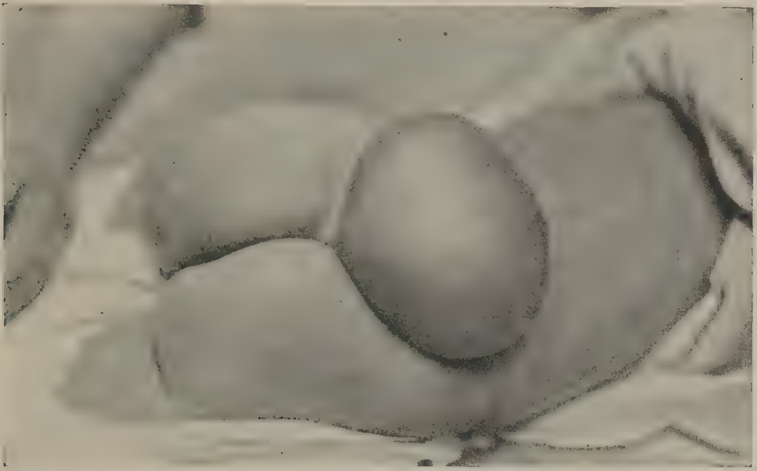

FIG. 37.—Spina bifida, meningocele.

Myelomeningocele or Meningomyelocele.—This is the most frequent and, at the same time, gravest type. The deformed cord is attached to the center of the hernial protrusion and from it nerve fibers pass down into the canal. The diagnosis can usually be made by the three zones described by V. Recklinghausen.* *Zona dermatica, zona epithelioserosa and area medullovasculosa.* In the center of the tumor mass is the area medullovasculosa, this looks like red juicy granulation tissue and bleeds on the slightest touch; outside of this area is the zona epithelioserosa which is composed of a thin translucent layer with numerous blood-vessels in it; outside of this area is the zona dermatica which is thickened skin often with a considerable growth of fine hair on it and numerous telangiectatic vessels; as it approaches the next zone the skin becomes thinner and takes on more the appearance of the cord membranes.

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* v. Recklinghausen, F., Untersuchungen über die Spina Bifida, Virchow's Archiv., cv (1886), p. 243.

Syringomyelocele or Myelocystocele.—In this type the central canal of the cord is greatly dilated and the wall of the hernial protrusion is composed of thinned out skin, dura and cord substance. It is usually covered with epidermis and therefore offers a slightly better prognosis.

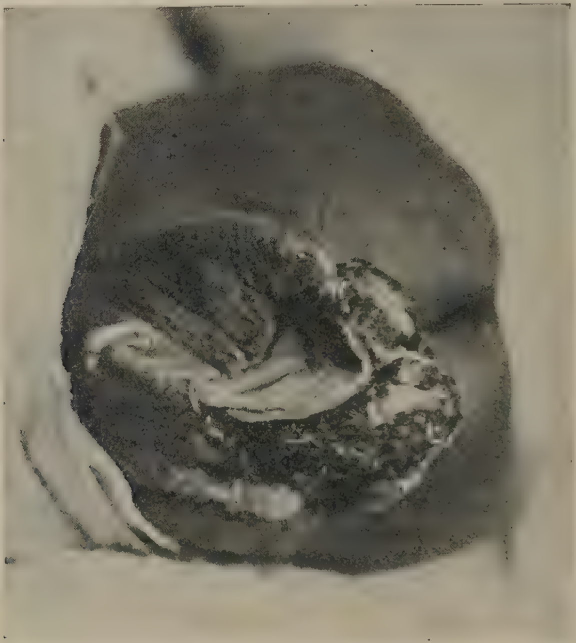

Fig. 38.—Spina bifida ruptured at birth.

Spina bifida occurs about once in every 1000 births. The mortality is very high, about 80 per cent. die in the first year and of the remaining 20 per cent. at least one-half succumb in the next few years. There are no very large statistics indicating how this mortality is affected by operative procedure but certain it is that the results are not very encouraging. I have had 25 cases with 14 deaths and of those that recovered some developed hydrocephalus. A considerable number of my cases have had perforations of the sac when they were admitted and this makes the operative risk very much greater since the danger of infection is added to the other operative risks (Fig. 38).

Diagnosis.—Whereas the diagnosis of spina bifida is made with little difficulty, the determination of the type of spina bifida is not so easy. In addition to the appearance on inspection, transillumination and determining the extent of involvement of the nervous system may help to establish the diagnosis.

In meningoceles the extremities and rectal sphincter are intact. If there are paralyses the spina bifida is more probably a myelomeningocele or syringomyelocele.

Treatment.—Authorities differ greatly both as to the proper time for operation and what the contra-indications are. Many surgeons take the attitude that one should wait until the child is a year old. I cannot share this view for if this policy were followed some cases that might be saved may be lost nor do I believe because a child may develop hydrocephalus one should desist from operation. If hydrocephalus develops that will have to be dealt with separately. It seems to me the only absolute contra-indications are: (1) A ruptured spina bifida; (2) deformities or complete paralysis of the legs. I cannot share the view of Hildebrand* that it is better to let a child die than let it grow up wearing braces. Certainly the wearing of braces is unfortunate but if the mentality of the individual is normal he may become a useful citizen in spite of his deformity. No physician or surgeon has the right, as I see it, to be a self-appointed lord high executioner and determine who shall be allowed to live and who shall not. It must ever be the purpose of the medical man to try to save life.

Of the various methods of treatment the only promising one is surgical. The injection of irritants to obliterate the sac is never to be undertaken. It neither relieves the local condition nor prevents the recurrence of the process. The sac is to be dissected out, the contents to be reduced into the canal and the dura to be carefully closed, serosa being sewed to serosa. The defect in the spine may then be covered with a flap of fascia from the erector spinæ muscle or by bone grafts as suggested by Albee† and first practiced by Trout.‡ Personally I have always felt it more desirable to close the defect so that there was some "give" to the closure than to have it closed as firmly as it would be by a bone graft. I have thought that this might make the formation of a subsequent hydrocephalus a little less probable.

Surgery of the Spinal Roots.—There are three conditions for which operations have been undertaken on the spinal roots. Pain, usually from malignant disease, the crises of tabes and spasticity. The first two of these are not conditions that occur in children and will not be discussed here. Spasticity, however, is a condition which occurs very frequently in children and certain of these cases may be relieved by surgical procedures carried out on the spinal roots.

The diseases variously spoken of as Little's disease, spastic diplegia and quadreplegia, are all characterized by intense spasticity of the extremities with pathological reflexes but no sensory changes. These conditions are natal or prenatal in origin. Hemorrhage and anemia during labor are probably the principal factors. Förster* advanced the theory that in these cases the spasticity is due to the blocking of the cortico-spinal pathway so that the inhibitory control of the brain is lost. As a result of this every sensory stimulus acts on the muscles and throws them into a state of tonic contraction—spasticity. He reasoned that if some of these sensory pathways could be removed the muscles would become flaccid. His theory has been contradicted but the undoubted fact remains that the cutting of sensory roots reduces the spasticity most remarkably.

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* Hildebrand, O., Die Behandlung der Spina Bifida, Arch. f. Klin. Chirurg., Berlin, xlvi (1893), p. 200.

† Albee, F. H., Original surgical uses of the bone graft, Surg. Gyn. and Obst., xviii (1914), p. 699.

‡ Trout, H. H., Spina bifida: tibial transplant father to child, Surg. Gyn. and Obst., Chicago, xx (1915), p. 523.

There are certain points that have to be carefully weighed before this operation is undertaken. A certain number of these spastic children are mentally deficient. Förster has pointed out that these children improve mentally after their spasticity is relieved and that has been my experience also but there must be some intelligence to work with, therefore the surgeon must determine beforehand how much coöperation he may expect from the child. In the second place, one must be certain that the patient has some voluntary control over his muscles so that when they become less spastic the patient will learn to use them. This is at times a difficult point to determine, especially if the spasticity is extreme. If the spasticity is due to a cerebral lesion I believe this voluntary control is always present. It is only when the lesion is spinal that all cerebral control is cut off, and in such cases cutting the posterior roots merely changes a spastic paralysis into a flaccid one. If the mentality is adequate and there is voluntary control of the muscles the operation may be performed. It is an operation that must not be undertaken lightly as it is a formidable one though not nearly so much of an undertaking in children as in adults since the lumbar spine lies much more superficially in children. I have always cut the roots at their exits as advocated by Tietze.† I do not believe the method used by Taylor‡ of cutting root fibers as they come off the lumbar enlargement is as good a procedure. I have always selected my roots depending on which muscles were most involved. As a rule I have cut the roots that Förster designated and which he believed afforded the best results, namely, Lumbar 2, 4, 5 and Sacral 2.

The operation can practically always be done in one stage. In young children a warm table should be used. I have done the operation *about* 18 *to* 20 times§ with one death so that it may be considered a fairly safe procedure. The aftertreatment of these cases is all important. Reeducation and massage must be employed consci-

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[*] Förster, O., Ueber eine neue operative Methode der Behandlung spasticher Lähmungen mittels Resektion hinterer Rückenmarkswurzeler, Ztschr. f. Orthop. Chir. Stuttg., xxii (1908), p. 203–223.

[†] Tietze, A., Die Technik der Förstereschen operation, Mitt. a.d. Grenzgeb. d. Med. u. Chir., xx, Jena (1909), pp. 559–564.

[‡] Clarke, L. P. and Taylor, A. S., The new treatment of spastic paralysis by resection of the posterior spinal nerve roots, New York Med. Journal, xci (1910), pp. 215–220.

[§] Sachs, E., A review of the operative results in ninety-four neurosurgical cases, St. Paul Med. Journ. (June, 1917).

entiously and for a long time. It sometimes is necessary to do some tendon lengthenings or tenotomies in addition to the root section. If these cases are treated by the neurologic surgeon and orthopedist together, the best results will be obtained.

Spinal Cord Tumors.—Spinal tumors are not as common as brain tumors but not by any means extremely rare. We shall consider here all types of pathologic processes that cause compression of the cord irrespective of whether they are true tumors or not for they all present very similar clinical pictures.

In children spinal tumors (neoplasms) are rarer than in adults. Frazier* reports 28 cases occurring in individuals from 1 to 20 out of a total of 330 cases. It is generally said that tuberculomas of the cord are more common in children than in adults. All processes in the spinal canal produce symptoms as a result of involvement of the cord. The process may be in the bone, in the spinal canal but extradural, intradural or intramedullary. The extra and intra dural tumors are as a rule dural tumors growing from the meninges and are the most favorable cases for permanent relief or complete cure. The tumors as a rule grow slowly and at first cause such obscure symptoms that the diagnosis may not be made for some time. Any type of tumor of endothelial or mesodermal origin may develop in the spinal canal. Primary carcinomas do not occur. Fibromas, lipomas, hemangiomas, all have been observed at operation or autopsy. Of the malignant tumors that have been described, sarcomata are said to be the most common, but I have the impression that just as in the case of brain tumors many of the older cases were really cases of gliomata that could not be differentiated as they can be today with special stains. There are several types of tumors which occur quite characteristically in the spinal cord.

Endothelioma, particularly the type known as **psammoma.** These are benign tumors growing from the endothelial cells in the dura and have peculiar calcified nodules scattered through the tumor that have the calcium deposited in concentric rings.

Glioma, though by far the most frequent tumor in the brain, is rare in the cord. They grow more rapidly than any other spinal tumor and, though infiltrating in character, never metastasize. They cannot therefore be called a true malignant tumor. Even if seen early, however, the prognosis is bad for the manipulation necessary to remove them is likely to cause permanent damage to the cord no matter how gentle the surgeon is.

Tuberculosis not infrequently affects the cord and meninges. It may affect the cord as a focal process, a true tuberculoma, or as a tuberculous abscess the result of a Pott's disease outside of the dura causing a compression myelitis. The abscess in these cases lies in front of the cord directly connected with the focus in the bone which always begins in the body of the vertebra.

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* Frazier, C. H., "Surgery of the spine and spinal cord." Appleton, New York (1918).

Gumma may occur in children but, since it follows acquired syphilis, is extremely rare and I find no record of such a case. The condition is not surgical.

The **symptoms and signs** of spinal tumor are of five kinds:

1. *Subjective Sensations.*—Tingling, formication or numbness.

2. *Objective Changes in Sensation.*—Hypesthesia or anesthesia—diminution or loss of the sensation of touch, which is usually tested with a wisp of cotton wool. Hypalgesia or analgesia—diminution or loss of the sensation of pain as tested by a pin. Thermhypesthesia or Thermanesthesia—diminution or loss of temperature sense.

3. *Paresis or paralysis* of muscle groups. The paralysis is of the central or spastic type. The muscles innervated at the site of the lesion may be flaccid.

4. *Changes in the Reflexes.*—Those reflexes are increased whose centers lie below the lesion while those whose centers are above the lesion are normal and if any centers happen to lie just at the level of the lesion the reflex originating from that segment will be lost.

5. *Pain.*—Root pain is a symptom that is of great value when present. It used to be believed that this symptom was essential to make the diagnosis of a spinal tumor but as clinical evidence has accumulated we have learned that this was not essential. It was then believed that pain was characteristic of dural tumors as distinguished from tumors growing in the substance of the cord. We know now, however, that dural tumors may frequently be unassociated with pain. This is particularly true of tumors in the dorsal region where the segments are quite long so that a tumor of considerable size may develop there without involving the posterior roots. If the roots are pressed upon the patients suffer very intense pain.

The determination of the level of a lesion requires very careful, often repeated, sensory examinations as well as a study of any paralyses and the reflexes. Though the paralyses may be enough to establish the level of the lesion the sensory examination much more frequently must be depended upon for this purpose. For this reason the diagnosis in children in the first decade becomes very difficult and in children below the age of five it is practically impossible unless there is a complete anesthesia. I have seen a child of 18 months in whom a level could be established because the anesthesia was complete but this is unusual. The charts (Fig. 39) give the innervation of various skin segments of the body quite accurately. There are other schemata which are used and these vary but slightly in detail.

In determining at which segment a lesion lies and where in reference to the spinous processes it is located, several anatomical and physiologic facts must be kept in mind. Sherrington has shown that no segment of the skin is supplied by one segment of the cord but that at least the adjacent segments, one above and one below, send fibers to each segment of skin. Thus the eighth skin segment receives fibers also from the seventh and ninth segments, therefore to get a total anesthesia of the eighth skin segment, the seventh must be involved, but to involve the seventh segment and throw it out of function the pressure must be in the upper portion of that segment or in the lower portion of the segment just above, namely the sixth. The spinal cord at birth extends the entire length of the spinal canal, as the child grows the spine grows more rapidly than the cord so that when full growth has been attained the cord ends opposite the first or second lumbar vertebra. Consequently the older the child, the higher the cord lies in its relation to the vertebræ. After the age of about eight the cord may be assumed to have the same relation it has in the adult. Reid* has made an elaborate study of this relationship. Chipault† has given the following directions for determining the proper level. In the cervical region the segment of the cord lies one higher than the corresponding vertebra. In the upper thoracic it lies two higher; in the lower thoracic it lies three higher. The last three lumbar segments lie opposite the eleventh thoracic spine and the sacral segments lie opposite the twelfth thoracic spine. Therefore if there were an anesthesia of the eighth skin segment the lesion would be in the seventh segment which lies under the

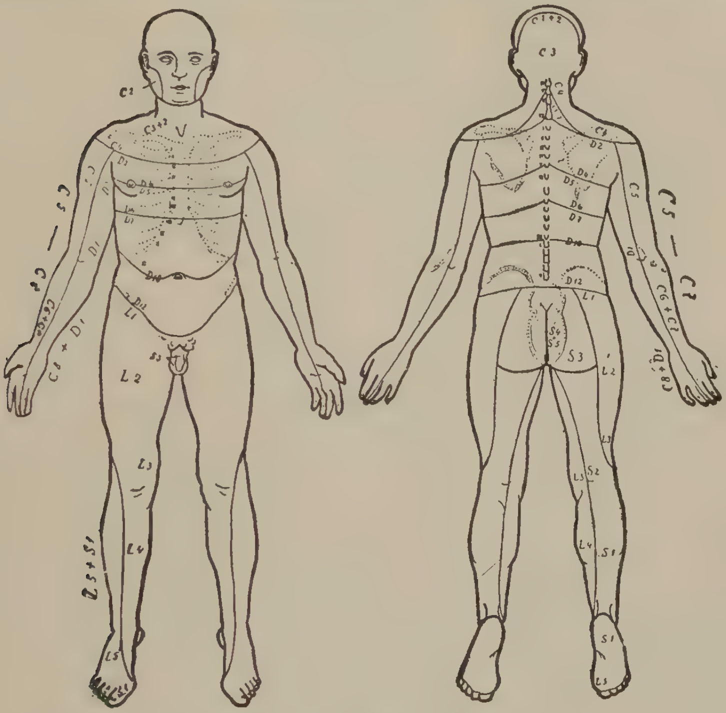

Fig. 39.—Seiffert charts.

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* Reid, R. W., Relations between the superficial origins of the spinal nerves and the spinous processes, Journ. Anat. and Physiol., xxiii (1889).

† Chipault, "Rapports des apophyses Epineuses avec la moelle." Thése de Paris (1893).

fourth spinous process. In addition to the changes in sensation mentioned above, occasionally an area of hyperesthesia marks the upper level of the lesion.

Treatment.—The treatment should always be surgical. Laminectomy is a very safe procedure and attended by a small mortality. If carried out properly there should be extremely little hemorrhage. The loss of cerebrospinal fluid which is inevitable when the dura is opened does no harm whatever. Very important, in my experience, it is to prevent any bleeding into the subdural or subarachnoid space. If this occurs the postoperative course is characterized by much pain, therefore whenever possible all the bone work should be completed before the dura is opened. If the primary incision is not big enough more spinous processes can be removed with care without in any way endangering the cord. Surgeons differ as to the best site for the incision. Frazier advocates making the incision from that lamina which lies over the lowest segment represented in the upper limit of the zone of anesthesia and carrying it upward. This may be a wise procedure since the great tendency is not to go high enough. Most surgeons make the probable site of the tumor the center of their field. I usually remove two spinous processes and then by the presence or absence of pulsation of the dura extend my incision upward or downward. As a rule the dura does not pulsate below the site of the lesion, but this is not an infallible rule.

There has been some discussion about operating in one stage or in two. I think there can be no question that the one stage is preferable except in the rare cases of intramedullary tumor where the extrusion operation of Elsberg* is used. This consists in incising the cord over the tumor at the first operation and then letting the tumor extrude itself and removing it at a second operation. Since most intramedullary tumors are malignant, this operation has a very limited application.

Trauma.—When the spine suffers a trauma, the injury to the bony structure is of comparatively little importance, the prime concern is what injury has the cord and its roots suffered and what harm may be produced later by the deformity.

The milder injuries, such as sprains and contusions of the spine, need no consideration here. They require no surgical procedure and clear up quite promptly with rest in bed and sometimes with the support of a plaster cast. It is of course important to take an *x*-ray in every case so as not to confuse a fracture or dislocation with the milder conditions.

Dislocations, Fractures and Fracture-dislocations.—As a result of one of these injuries, the vessels in the canal may be ruptured and a hemorrhage may take place into the canal or there may be extravasation into the subcutaneous tissue. There may be injury to the nerve roots or there may be compression of the cord itself. These three types of lesions may be produced by any one of these three injuries.

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* Elsberg, C. A., The surgical treatment of intramedullary affections of the spinal cord, Surg. Gyn. and Obst., xviii (1914), p. 170.

Dislocations.—Pure dislocations of the vertebræ are rather uncommon as they are so often associated with fractures. They may be unilateral or bilateral. The deformity when it occurs in the cervical region may help in the diagnosis, though very similar deformities may be observed in fracture dislocations. A good *x*-ray plate and still better stereoscopic plates are invaluable aids in arriving at a diagnosis. In fact nowadays a plate should always be taken. A blow on the head, diving into shallow water, falling and striking the head may all bring about such an injury. The dislocations of the axis upon the atlas may be backward or forward and these may be bilateral or unilateral. The deformity is quite characteristic. Examination of the pharynx with the palpating finger will often give the clue to the condition. The head may be held forward and rotation to either side may be impossible. If the dislocation is unilateral, motion may be limited to the side of the lesion. If the odontoid is fractured, still greater care must be exercised in handling the case as manipulation may result in instant death. I have seen a patient with a fracture dislocation of this sort instantly killed by the manipulation of an osteopath. Reduction should only be undertaken after careful *x*-ray studies. If a fracture is associated, absolute rest and immobilization are necessary. After several weeks the patient may be allowed to sit up with a support and if there is no increase of his symptoms he may be allowed to walk. In dislocations in the lower cervical and dorsal spine the spinous processes do not lie in the median line and at the site of dislocation there may be a depression or prominence depending on whether the dislocation is forward or backward. It is quite impossible from the examination alone to determine that there is only a dislocation. The same may be said of a lumbar dislocation.

Treatment.—If the dislocation is uncomplicated by a fracture, the indication is to attempt a reduction. This should be done under general anesthesia both on account of the pain and to relax the muscles. The reduction is dangerous and the family should be made fully aware of this fact. Even if there are no cord symptoms, most authorities are agreed that it should be attempted to forestall an increase of the deformity which might produce cord symptoms. It is usually recommended to do a closed reduction but Frazier has very wisely drawn attention to the great value of a laminectomy in cases where there are cord symptoms. In these cases the cord may not be pressed upon by the bone but be compressed by a hematoma. It certainly has seemed to me the proper procedure if carried out by a surgeon experienced in laminectomy. I believe in applying the same rule to dislocations that I have advocated in fractures, namely, immediate operation. In old dislocation of more than two weeks standing closed reduction is extremely dangerous and if there are cord symptoms an exploratory laminectomy should be performed. Only a spinal decompression may be done but this may be quite enough to relieve the pressure on the cord.

Fractures of the spine may involve the spinous process, the articular processes, or the transverse processes of the body. There is no need of a lengthy discussion here as to the causes of these different types of fractures. Direct trauma is the usual cause. To make an accurate diagnosis an *x*-ray plate is essential.

The type of treatment depends on whether or not there is evidence of cord injury. If the cord is injured, operation should be advised. If there are no cord symptoms, immobilization with a jacket or corset are to be employed. In general, it may be said that fractures of the spine unaccompanied by dislocation have a more favorable prognosis and have fewer symptoms than either dislocations or fracture-dislocations.

Fracture-dislocation may be produced by direct or indirect violence. Some exaggeration of one of the normal movements of the spinal column as a rule brings it on. This is generally speaking the gravest type of injury, for either the dislocation or the fracture may compress the cord. The deformity may not be very great. There may be faulty alignment of the spinous processes, a depression or a kyphos, but in some cases, especially fat individuals, it may be difficult to discover anything abnormal. Examination should be most careful; no attempt should be made to elicit crepitus and all manipulations and movements of the patient should be confined to the most necessary ones.

The treatment depends upon the extent of the cord symptoms. If there is evidence of compression, operation should be advised. Some difference of opinion exists as to the best course to follow if the symptoms point to a complete transverse lesion of the cord. This will be taken up below.

We come now to the various types of injuries to the cord that may occur with dislocation, fracture or fracture dislocation.

Concussion of the Spine.—On page 118 of this volume under the caption "Concussion of the Brain" I have expressed my views on this subject. What has been said there holds true of concussion of the spine. The well-meaning but faulty observations of Erichsen in 1875 have started a flood of literature and litigation. Frazier* in his book, p. 372, summarizes the subject very well when he says "We, therefore, hold to the opinion that where there is injury to the spinal column without a bony lesion but with subsequent alteration in the spinal cord it were well to class such a case as contusion and to eliminate the term 'concussion' as applied to the spinal cord altogether." I want to repeat what was said in discussing concussion on page 118. There is no clinical picture characteristic of concussion of the cord and therefore there can be no sound reliable pathology.

Hematorachis and Hematomyelia.—Hemorrhage may occur into the subdural or subarachnoid space or into the cord itself. If it is in the subdural space—hematorachis—the picture may be compared to a middle meningeal hemorrhage, that is, slowly increasing paralysis is very suggestive of such a condition. There may be pain from compression of the roots but this is in no way essential. Hematorachis may occur occasionally with other conditions than fracture—tetanus, infectious diseases, or meningitis and at times in a fractured skull in which the blood has flowed into the spinal canal. The condition may be suspected but it is doubtful if a positive diagnosis can ever be made. Lumbar puncture may show crenated blood cells and this may strengthen the suspicion, though the same picture throughout might be seen in a case of contusion and laceration of the cord. I have once seen a case where the slowly increasing symptoms in the course of four to six hours made it possible to make a positive diagnosis which was proved at operation.

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*Frazier, C. H. and Allen, A. R., "Surgery of the Spine and Spinal Cord." With the collaboration of Alfred Reginald Allen, N. Y. and London, Appleton, 1918.

In *Hematomyelia* the hemorrhage is in the substance of the cord and may occur independently of an injury to the spinal column. The hemorrhage may be minute or very large extending over several segments of the cord. The symptoms vary tremendously depending on the level of the lesion as well as its location in the cord. If it occupies the central canal the picture may be that of a syringomyelia—dissociation of sensation, loss of pain and temperature with retained tactile sense. In other cases there may be a Brown-Séquard syndrome—loss of motor power on one side and sensory disturbances on the other side.

The treatment here depends on the severity of the picture. I believe that if some of these cases are operated early and the blood clot is evacuated a far better result may be obtained than if they are left alone for in that case they always develop a syringomyelic cavity which is a hopeless condition.

Contusion and Laceration.—These two conditions must be considered together for the latter is but the aggravation of the former and all gradations may be found from a small ecchymosis to a complete disintegration of the entire cord. There may be a great deal of hemorrhage or there may be mostly traumatized cord tissue. The monumental studies of Reginald Allen* have advanced our knowledge of this phase of the subject tremendously. He not only showed the various degrees of these lesions but pointed out that the hemorrhage and edema that accompanied these lesions were responsible for many of the symptoms. He then went a step farther and proved on animals that if the cord were incised soon after the injury to relieve the tension produced by the edema and hemorrhage, far more would recover and with fewer residual paralytic symptoms. This work should receive far more consideration than it has thus far. I have planned my treatment of these cases on Allen's findings and have for some years advocated and practiced immediate exploration of every case of spinal fracture with *marked cord symptoms.* I realize that this is generally considered radical but my reasons are these. If the cord has been completely destroyed, exploration has done no harm. If the picture of a complete transverse lesion is due to hemorrhage and edema, early operation will save some of these cases. Exploratory laminectomy when done with care is quite safe so that the possible benefit far outweighs the operative risk.

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* Allen, A. R., Remarks on the histopathological changes in the spinal cord due to impact, Journ. of Nerv. and Ment. Dis., Lancaster, Pa., xli (1914), p. 141.

Symptoms of Partial and Complete Cord Lesions.—The determination of the level of the lesion is important, though with the history of trauma and either the physiological and *x*-ray findings to help us in knowing which vertebra has been injured, that phase of the problem is usually far simpler than in the case of spinal tumors where we have only the neurologic findings on which to base our diagnosis. Of far greater importance is this question. Is there a complete or partial lesion of the cord? It is surprising how much beating around the bush there is on this question. If there is any sensation below the level of the lesion, if some of the reflexes are retained or there still are some movements, there is no question whatever that some of the tracts in the cord are intact. The great majority of such cases may be safely left alone for they recover remarkably and at times completely. If immediately after the accident, below a certain level all reflexes are gone, all sensation lost and complete motor power gone, does that mean that there is a complete break in the cord; if not, by what symptoms or signs may a complete break in the cord be diagnosed?

This is the vital question which must be answered. Up to the present time *there exists no method of determining whether a lesion of the cord is complete or partial if all reflexes, motor and sensory functions are absent.*

The level of some lesions may be readily recognized by characteristic attitudes or other signs. Thus if the sixth cervical segment is affected, the arms and forearms will be flexed due to the paralyses of all the extensor muscles (Thorburn).* When the lesion is at the seventh segment, an equally characteristic picture is seen. Below this level the outline of the sensory loss will enable the surgeon to determine the level of the lesion.

Treatment.—I have expressed my views on this subject above. It may be well, however, to summarize them once more. Early operation where the picture of a complete transverse lesion is present is the method of choice since it holds out hope of recovery to those cases which have nervous tissue that still can functionate.

Gunshot Wounds.—Just a word needs to be added here on this subject. In children the only wounds of this sort are the accidental ones of civil life. The rules that govern these cases are quite similar to those outlined for spinal injuries in the preceding pages. We have here the added complication of an external injury. Cleaning of the wound along the lines of a "debridement" are indicated. The spinal injury is to be treated along the same lines as indicated above, with this difference; whereas a partial cord lesion might be left alone in the case of a fracture, in the case of a gunshot wound operation should be undertaken to avoid if possible a later infection, and the bullet if it lies in the canal should be removed. If the bullet is buried in the bone where its removal would involve much manipulation it might be left in situ.

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* Thorburn, W., Lectures on the surgery of the spinal cord, Brit. Med. Jour., i (1894), p. 1345.

Inflammations.—The various types of inflammation will not be discussed here, merely those conditions will be briefly referred to which may be relieved by surgical measures. These are:

Pachymeningitis Cervicalis Hypertrophica (Oppenheim).*—This condition consists of an inflammatory thickening of the membranes about the cervical cord. There is no specific organism responsible for it. It may occur following a hemorrhage in that region. It occurs in older individuals though it may occasionally be found in children. The symptoms are those of compression of the cervical cord. The disease is slowly progressive and resembles the clinical picture produced by a cord tumor in that region. Operation and severing the adhesions has afforded relief in a number of instances.

Tuberculosis.—In most cases surgical interference is not to be thought of but occasionally, if the usual orthopedic measures of traction and extension do not help, evacuation of an extradural abscess has been attempted, followed by closure of the wound without drainage, just as in recent years has been done with psoas abscesses. The procedure is not very satisfactory and is only to be considered if all other measures fail.

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* Oppenheim, H., "Lehrbuch der Nervenkrankheiten." Karger, Berlin (1905).

CHAPTER CLXX

CHOREA

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Synonyms.—Chorea Minor, Sydenham's Chorea, St. Vitus's Dance.

Historical.—The terms chorea and St. Vitus dance have in the past been loosely applied to a variety of conditions which are now recognized as being widely different in their etiology and symptomatology. To mention only one of these groups, it is interesting to note that during the Middle Ages, Continental Europe was the scene of numerous widespread "epidemics" of a peculiar form of "dancing mania" or "dancing disease" to which the name of "St. John's dance" or "St. Vitus's" dance was applied. These, however, are now known to have been for the most part manifestations of hysteria, and bore no relation to the disease which forms the subject of this chapter. It became customary for persons afflicted with this hysterical malady to seek relief by making pilgrimages to the shrines of certain saints, and accordingly in different parts of the country the disease was referred to as St. Anthony's dance, St. John's dance, or St. Vitus's dance. In the sixteenth century Paracelsus gave the name Chorea Sancti Viti* (chorea, meaning "dance") to this epidemic dancing disorder, and the latter term was generally used in referring to it thereafter.

The name St. Vitus's dance, however, soon came to be applied to another form of disorder in which the movements consisted of more or less incoördinated, purposeless jerkings and twitchings, rather than dancing movements. It was this latter type which Thomas Sydenham[3] described in England in 1686 as St. Vitus's dance, making no mention of epidemic dancing manias, but unfortunately using the same name. Indeed it seems probable that the latter, at least in the form which had swept through Germany, had not appeared in England at this time. Sydenham's description, although meager, contained many of the essential features of chorea as we know it today.* Thus he noted especially that it was a disease of childhood, which produced irregular, incoördinated twitchings, often limited to one side of the body. Great confusion existed in the subsequent literature, concerning "Chorea Anglorum" and "Chorea Germanorum," until Wichmann,[4] in 1801, emphasized the fact that the English conception of St. Vitus's dance was entirely different from the German, and suggested that the English species was a lesser or milder variety of the German type (hence the terminology "chorea minor" and "chorea major"). During the following century it became customary to apply the generic term chorea to almost any disorder characterized by purposeless movements or twitchings, and dozens of varieties of chorea were mentioned in the literature. With greater precision in the study of disease, came a tendency to drop the name St. Vitus's dance altogether and to limit the application of the term chorea to the disease first described by Sydenham. The latter is also used, however, in the descriptive names of several other disease, as for example *chronic progressive chorea* (*Huntington's chorea*), *chorea electrica*, etc.

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* The story of how St. Vitus became the patron saint of those afflicted with the dancing plague is of interest. Vitus was a Sicilian youth who suffered martyrdom at the time of the persecution of the Christians under Diocletian in A.D. 303. He was said to have been buried at Florence, and it was not long before the miraculous powers of the saint were proclaimed throughout Italy. In the eighth century his body was removed to the church of St. Denys in France, and in the year 836, to Corvey, each such removal serving to stimulate interest in the saint and to spread his fame still farther. A later legend relates that just before St. Vitus bent his neck to the sword, he prayed to God that he might protect from the dancing mania all those who should solemnize the day of his commemoration, and that thereupon a voice from Heaven was heard crying "Vitus, thy prayer is granted!"[1, 2]. Shrines to his memory were erected in many places, some of the more famous ones being in Zabern, Ulm, and Ravensburg.

Etiology.—Predisposing Causes.—*Age.*—Chorea is rare in the first few years of life, the vast majority of cases developing between the ages of 5 and 15 years with the greatest incidence between the seventh and thirteenth years. Thus in M. Allen Starr's[5] large series of 2239 cases, 1600 or 71 per cent. occurred between the fifth and fifteenth years and only 36 (less than 2 per cent.) before the fifth year. Thayer,[6] in an analysis of 808 cases, found 2.5 per cent. under five years and 84.5 per cent. between 5 and 15 years. Of Osler's[7] 522 cases, 440 (84.4 per cent.) were between 5 and 15 years, and 33 (6.4 per cent.) under five. Mackenzie reporting the 439 cases of the Collective Investigation Committee of the British Medical Association,[8] found 1.4 per cent. under five years and over 77 per cent. between 5 and 15 years.

Though the disease is extremely rare before the third year, it is seen occasionally even in infancy. Various writers from time to time have even reported instances of supposed congenital chorea, and V. Dupuy[9] has collected 29 such cases from the literature, but there would seem to be considerable doubt as to the identity of these cases with true chorea. The youngest child in Holt and Howland's[10] 146 cases was four years, in Abt and Levinson's[11] series of 226 cases 3½ years, and in Mackenzie's[8] three years.

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* Sydenham's description, as given in the third edition (1753) of Swan's translation from the original Latin is as follows: "This disorder is a kind of convulsion, which seizes children of both sexes, from the tenth to the fourteenth year; it manifests itself by a halting, or unsteadiness of one of the legs, which the patient draws after him like an idiot. If the hand of the same side be applied to the breast, or any other part of the body, the child can't keep it a moment in the same posture, but it will be drawn into a different one by a convulsion, notwithstanding all his efforts to the contrary. Before a child who hath this disorder can get a glass or cup to his mouth, he useth abundance of odd gestures, for he does not bring it in a straight line thereto, but his hand being drawn sideways by the spasm, he moves it backwards and forwards, till at length the glass accidentally coming nearer his lips, he throws the liquor hastily into his mouth and swallows it greedily, as if he meant to divert the spectators."

Sex.—All observers agree that chorea is much more prevalent among females than males, the proportion being given by Starr[5] (2239 cases) as 2:1, by Gower's[12] (compilations of 1365 cases) as 3:1, Mackenzie[8] (439) as 2.8:1, Abt and Levinson[11] (226 cases) 2:1, Osler[7] (551 cases) slightly over 2:1, Wollenberg[13] compiled statistics on 3595 reported cases of chorea, and found 2481 (69 per cent.) in females and 1114 (30.9 per cent.) in males, or a ratio of 2.2:1.

Heredity.—The influence of heredity is uncertain. Among the parents of choreic children, Starr[5] found 7 per cent. had had the disease in early life. The occurrence of two or more cases in the same family is somewhat less unusual, Osler[7] reporting 80 such instances in 554 cases. It is quite common, on the other hand, to find a history of "nervousness" or various neurotic tendencies in the parents, which often extends to the patient as well. Finally, if one includes in a study of the heredity familial rheumatism as well as familial chorea, as some writers insist should be done, the percentage of positives rises considerably higher. Wollenberg,[13] for example, adding his cases to those of Money and Sturges, found 68 of 426 cases (16 per cent.) gave a history of a rheumatic taint. Garrod,[14] broadening the conception of rheumatism somewhat, found 38.8 per cent. positive. The relation of chorea to rheumatism will be considered more fully a little later.

Race and Climate.—The disease is said to be less common in hot climates, and is distinctly rare among negroes and Indians. Griffith[15] states that he has seen only two cases in negroes, and Osler[7] reports five negroes among 175 cases of chorea at the Johns Hopkins Hospital. Thayer,[6] however, in a later report from the same institution, found 3 per cent. of the chorea patients (25 of 808 cases) were negroes, although the colored race made up 12 per cent. of the total dispensary and hospital attendance. At the Washington University Dispensary in St. Louis, there were seven negroes among 220 children with chorea.

Season.—By most writers, the season of the year is thought to have a distinct influence on the occurrence of chorea, the larger number of cases occurring in the spring months. Lewis,[16] in a collection of 1383 cases in Boston and Philadelphia, found by far the greatest number occurred in March, April and May, the incidence being lowest in November. Haven,[17] in 146 cases, found 38.3 per cent. occurred in the spring, 27.6 per cent. in the winter, 20.5 per cent. in the summer and 13 per cent. in the fall. Koch,[18] in 100 cases, found 29 per cent. began in the first three months of the year, 21 per cent. in the next three months, 13 per cent. in the third quarter, and 37 per cent. in the fourth. Abt and Levinson,[11] however, report that season did not play a constant rôle in their series of 226 cases. The greatest number occurred in January and the smallest number in October. Our own cases (220), divided into quarterly periods, showed 69 the first quarter, 53 the second, 62 the third, and 36 the fourth.

Inciting Causes.—Various *psychic influences* appear at times to have a decided effect on the onset of chorea. *Fright, worry,* sudden *mental shock* or great *emotional strain* are frequently mentioned in the history as having preceded the choreic manifestations. *Overstudy,* or, the mental strain of school work is also a prolific source of trouble in this respect. Often, however, it will be found that certain of the milder mental and motor symptoms of the disease were actually present before the occurrence of the psychic trauma, but have been overlooked by the parents. *Imitation* is thought by many writers to be an occasional cause for the spread of chorea, particularly in institutions, producing localized "epidemics." Many such cases, instead of true chorea, are doubtless instances of hysteria or tic-like movements in neurotic children, for it is well known that such children frequently imitate other irregularities of movement which have no relation to chorea. Still[19] says with respect to this point "Amongst many hundreds of cases of chorea which have been under my observation in various institutions I have not seen a single instance of the spread of chorea from one child to another by imitation; and this in spite of the absence of any effort to screen off the choreic child from the observation of others." Numerous *reflex causes* have been mentioned as contributing to the onset of the disease, but their influence must be considered as extremely doubtful. Among those which have been suggested are intestinal parasites, beginning or delayed menstruation, ocular defects, etc. Errors of refraction frequently cause tic-like spasms of the ocular muscles which simulate, but are entirely distinct from true chorea. The *acute infectious diseases,* particularly scarlet and typhoid fevers, are sometimes followed by chorea, although the association of the two is not very striking. Milian[20] thinks there exists a very close relationship between *syphilis* and chorea but this seems to be quite definitely disproved by many other investigators. *Anemia* and *malnutrition* may be important contributing causes, although they are more apt to develop as later symptoms of the disease.

Relation of Chorea to Rheumatism.—It has long been recognized that there is an intimate relationship between chorea and rheumatism, but until the etiology of the two conditions is more definitely established, the precise nature of this association cannot be determined. According to Wicke[21] even as early as 1780 Stoll described several cases of rheumatism associated with chorea, and in 1810 Bouteille,[22] reviewing these cases, and 2 of Sauvage's, added another one of his own, and proposed the name "rheumatic chorea" for the condition. Similar observations were made by Copland[23] (1821), Thomson[24] (1831), Bright[25] (1839), Babington[26] (1842) and others. Bright, writing in 1838, emphasized the association of chorea and heart affections, adding that the connection of chorea with rheumatism had long been recognized. He states that "in the very excellent 'Syllabus or Outlines of

Lectures on the Practice of Medicine' published at Guy's Hospital, I found in the edition of 1802, rheumatism distinctly stated as one of the existing causes of chorea; and in later editions, as in that of 1820, I find it stated that 'chorea sometimes alternates with acute rheumatism, but through what organ or by what intervention this occurs is not conjectured.'' Similarly, Hughes,[27] who in 1846 published a "Digest of 100 cases of chorea,'' and in 1855 with E. B. Brown,[28] analyzed an additional 209 cases, and Seé[29] (1850), did much to initiate the many systematic and careful studies which have sprung up since this time concerning the association of these two conditions. Hughes and Brown stated that of 104 cases in which special inquires on these points were made, there were only 15 patients who were "both free from cardiac affections and had not suffered from a previous attack of rheumatism.''

These views met with considerable opposition on many sides, but found an able champion in Roger[30] (1866), who not only vigorously defended them, but expressed his belief that acute articular rheumatism, chorea and the accompanying heart affections were all parts of the same disease, and proposed a distinctive terminology which should make this point clear.

Probably one of the chief reasons for such differences of opinion as existed in the past, and exists in some quarters even today, is the confusion as to just what is meant by "rheumatism.'' If we include under this heading only those cases with sudden onset, high fever, and marked inflammatory joint involvement, undoubtedly the percentage of children with chorea who give a rheumatic history will be small. If, on the other hand, we consider, as most pediatricians do, that rheumatism in childhood runs a much less stormy course than in adults, and that frequently its manifestations are so slight as to be entirely over-looked or attributed for instance to "growing pains,'' then indeed the incidence of rheumatism in chorea is very much increased. Statistics on this point vary rather widely, probably for the same reason. Thus P. Meyer[31] gave the incidence as 9 per cent., Sturges[32] as 19 per cent., and Mackenzie[8] as 32.5 per cent. whereas Duckworth[33] claims that 85 per cent. of his cases gave a family or personal history of rheumatism. In Starr's[5] series of 2500 cases, which is one of the largest published, 26 per cent. gave a rheumatic history. Batten,[34] in a careful study of 115 cases of chorea, found that on admission 32.2 per cent. of the children gave a history of having had rheumatism, but when these same cases were followed for a period of six years thereafter, the incidence rose to 53 per cent. Finally, G. F. Still,[19] feels that we have much under-estimated the frequency of rheumatic manifestations, and asserts that he regards chorea as "just as good evidence of rheumatism as a gumma is of syphilis.'' However this may be, there has been a tendency recently to consider the higher figures (50 to 75 per cent.) as probably more nearly correct. In our series in St. Louis, 61 per cent. gave a history of having had a previous attack of rheumatism, 36 per cent. gave a rheumatic family history, and 83 per cent. had had tonsillitis.

Tonsillitis and Chorea.—The association of tonsillitis and pharyngitis with rheumatism and chorea often appears to be a very close one. Of 90 children in our series in which a history of this symptom was especially searched for, 75 had previously had one or more attacks of tonsillitis. Abt and Levinson,[11] however, in 135 cases found only one-third with such history. Nevertheless, by many clinicians, recurrent attacks of tonsillitis are considered presumptive evidence of a rheumatic tendency and the tonsils regarded as the portal of entry for the organisms of rheumatism.

Bacteriology.—The theory that chorea is an infectious disease, caused by a specific microörganism, has gained great prominence during recent years and has much to support it. The frequency with which endocarditis is found in chorea would in itself be strong presumptive evidence of its correctness. And if it is assumed that the same organism is capable of producing different lesions and different sets of symptoms under appropriate conditions, an easy explanation is afforded of the relationship between chorea, rheumatism, heart affections and tonsillitis.

Even though the infectious origin of chorea be conceded, the bacteriology of the disease is still unsettled. Numerous investigators have reported the finding of bacteria in the blood, spinal fluid, brain or meninges, but the lack of uniformity in the results has cast a certain amount of discredit upon all. One of the earliest of such studies was that by G. Pianese,[35] who in 1891 reported the isolation of a diplobacillus from the brain of a child with chorea. Animals inoculated with this organism developed muscular twitching and convulsions, and at autopsy the same bacillus was obtained from the central nervous system. These results, however, have never been confirmed. Among the many other writers who have reported occasional positive findings in cultures from the blood and tissues, may be mentioned Dana,[36] Westphal, Wassermann and Malkoff,[37] Poynton and Paine,[38] Richards,[39] Camisa,[40] Maragliano,[41] Heubner,[42] Donath,[43] Morse and Floyd,[44] Quigley,[45] Griffith[15] and others. The organisms found have in most instances belonged to the streptococcus or staphylococcus group, the former predominating. Quigley reports 21 cases with no active heart or joint symptoms, 10 of which yielded positive cultures from both blood and spinal fluid, and 14 in either one or the other. Eight of the strains isolated from the spinal fluid and nine from the blood were identical in their morphologic and cultural characteristics and seemed to belong to the same group of streptococci or diplococci frequently found in rheumatic fever and endocarditis. Poynton and Paine consider chorea a manifestation of rheumatism and they claim to have isolated from the blood in chorea a diplococcus similar to that found by them in cases of rheumatism.

In many instances, the bacteriologic findings lose in value from the fact that in addition to the chorea, there is present at the time of the blood culture, a complicating endocarditis. Positive findings in the acute stages of the latter disease are quite common, and though it is possible that both the chorea and the cardiac affections may be due to the same organism, the connecting link is missing. Furthermore the presence of endocarditis may be unsuspected at the time of the investigation as in the following example: A child was admitted to the St. Louis Children's Hospital with severe chorea and fever for which no other cause than the chorea itself was found, the heart being apparently normal. From the blood, a diplococcus was isolated which produced arthritis and endocarditis when injected into rabbits. Within a few days after the blood culture was taken, however, it became quite evident that the child's fever came from a complicating acute endocarditis, thus detracting somewhat from the interest of the bacteriologic findings.

To sum up, therefore, we may say that the theory that chorea is an infectious disease has gained a wide acceptance among clinicians, and there is much evidence to support it. Complete confirmation of this theory, however, is still lacking owing to the inconclusive nature of the bacteriologic findings.

Pathology.—The pathology of chorea has been the subject of much speculation and investigation, but very little is actually known about this complex subject. Opportunities for autopsy examination are very limited in chorea, because of the infrequency with which death occurs during the period of active symptoms. The mortality is low and fatalities which occur as the result of heart complications usually take place long after all other choreic manifestations have subsided. It is not surprising, therefore, that with the few exceptions to be noted below, the pathologic examination is usually negative. Indeed the very nature of the motor phenomena, which constitutes the most striking feature of chorea—the irregular, wide-spread distribution of the muscular twitching, and the transient nature of the affection—speaks more for a functional derangement than one with an anatomic basis.

Among the lesions which have from time to time been found at autopsy in chorea, the one which has perhaps received the greatest prominence is the presence of multiple capillary emboli in the brain. These, however, have not been found in all cases, and are believed to be due in all probability to a complicating endocarditis. Money[46,47] attempted to produce similar lesions in animals by injecting minute particles of foreign matter (e.g., starch granules, bismuth, etc.) into the carotid artery. Although certain of the animals so injected developed movements of the limbs, it is doubtful whether these were truly choreic in nature. Other vascvlar changes occurring in the brain and cord have also been met with occasionally, as for example intense hyperemia, perivascular infiltrations, vacuolizations of the tissues, and minute hemorrhages. Such lesions suggest an infectious process, and it is not surprising that bacteria, usually of the streptococcus group, have been isolated at times from the brain and blood stream, as mentioned in the preceding chapter. They lend support to the theory of the infectious nature of chorea, particularly those cases with rheumatic history or resulting heart affections.

In the absence then of any more uniform or definite anatomic picture, it is impossible to say more about the pathologic process than that from the symptoms it would appear to affect chiefly the central nervous system, producing signs of cortical irritation in the motor areas. That any changes which may occur in these localities are of a superficial and transitory nature with little structural change, is indicated by the fact that complete recovery within a period of several weeks to a few months is the rule in chorea.

Symptoms.—The onset is usually insidious. Occasionally, however, the motor symptoms appear to be precipitated by sudden fright or other psychic trauma. In such instances, careful questioning will often bring out the fact that choreic manifestations in reality existed before the supposed inciting cause, the latter producing a sudden exacerbation of a condition already present rather than the onset. In the majority of cases, the parents first notice that the child is irritable, restless, fidgety, nervous, sleeps poorly and cries very easily for no apparent cause. Next a certain awkwardness develops, which is shown by the child's frequently stumbling, dropping objects from the hands, spilling food, etc. Closer inspection at this time will usually reveal some slight twitching or jerking which may have completely escaped the notice of the family. If the hands are extended with the fingers widely separated, a fine tremor is usually noticeable, as well as occasional irregular twitching of the fingers. The fingers are almost invariably markedly hyperextended. The coarser movements may be brought under control of the will for a time, but if now the patient is asked to protrude the tongue also, not only will the tremor be found to involve this organ as well, but an intensification of all the movements elsewhere in the body commonly takes place. Often efforts at repression serve but to exaggerate the condition. It is however, characteristic of the milder forms of the disease that all movements cease during sleep. Headache, fatigue, slight pain in the limbs, loss of nutrition and the development of anemia are all frequent even in cases of only moderate severity.

Mild cases may never progress beyond this point. More commonly, however, there is a gradual accentuation of all the symptoms until the picture becomes so striking as not to be easily confused with any other condition. The child fidgets and squirms in its seat, the shoulders are shrugged, the arms are jerked about, the fingers twitch, and the victim apparently suffers the most intense embarrassment at the mother's recital of the train of symptoms. This impression of the patient's confusion is heightened by the frequent laughter without cause, to be followed a moment later by a flood of tears. Grimaces are frequent, but the face in repose takes on a vacant expression which is suggestive of mental deficiency. Speech too, is affected, the degree varying from a slight slurring and stumbling over words, or explosive speech, to a complete inability to articulate. Swift[48] has also described characteristic choreic changes in the voice, consisting of alterations in pitch and intensity. Complete loss of speech has been known to persist for over six months. In most cases difficulty in speech is due to the loss of control over tongue, lips, and palate, the larynx being less commonly involved. For the same reason the swallowing of food may become quite difficult. Involvement of the tongue and lips may be easily demonstrated by requesting the patient to thrust out the tongue and hold it out for inspection. Despite the obvious lack of control over these muscles, the tongue is practically never bitten.

The deficient muscular control causes an awkwardness which the child tries desperately to overcome. The result is over-action of the muscles on attempting any specific task, such as buttoning a coat or unlacing a shoe, and consciousness of this bungling of a simple task results in a further confusion of the patient. Frequently one side of the body is much more involved than the other (hemi-chorea) although some jerking can usually be detected even on the better side. Loss of power in the affected muscles is present in all of the severer cases, and may be so pronounced as to simulate paralysis. Such cases, in which paresis is a more prominent feature than jerking, are sometimes referred to as *paralytic chorea* or *chorea mollis*. When this weakness affects chiefly one limb or one side of the body, the resemblance to monoplegia or hemiplegia may be quite striking at first, until the presence of the typical twitching both here and on the less affected side, reveals the true nature of the condition. Again, loss of power in the extremities may be so marked as to make walking impossible and interfere seriously with all voluntary acts. It must be remembered in speaking of the different types of chorea that frequently one form merges insensibly into the other. Thus a mild chorea may be converted into a severe case with or without a marked disproportion in the involvement of the two sides, and similarly the wildest gesticulations may be superseded by chorea of the "paralytic" variety.

In all patients with moderately severe chorea, there is present some disturbance in the *mental state*. This may be manifested merely by fretfulness, lack of concentration and an emotional instability which causes the patient to burst into tears or laughter on the slightest provocation. Not uncommonly, however, the face takes on a vacuous expression, broken by frequent grimaces, which strongly suggests beginning imbecility or dementia. Frequently this demented look is heightened by retraction of the upper eye lid (Stellwag's sign). Occasionally, the psychic disturbance may be so marked and violent as to simulate acute mania. Rarely, hallucinations melancholia, and even stupor, have been reported. Such severe psychic reactions are distinctly less common in children than in young adults.

The *muscular movements* in chorea are very striking and characteristic. They are rapid, irregular, involuntary, and of wide excursion. They are not limited to one particular set of muscles, but affect widely separated groups in irregular order. Ordinarily, they are first noticed in the hands, but soon thereafter the facial muscles become involved and then the shoulders, lower extremities, and trunk, in irregular order. The intensity and amplitude of the movements varies from a slight twitching of the fingers and facial muscles to the wildest jerking about of extremities and trunk. In chorea of the severest type, the movements take on a frightful intensity (chorea insaniens). The patient threshes about in bed continuously, with a violence that promises severe bodily injury unless he be restrained in some manner. Utter helplessness, complete inability to talk and persistence of the movements even in sleep, are common in this form of the disease.

Hemi-chorea in which there is a marked disproportion between the involvement of the two sides of the body is, as mentioned, quite frequent. It occurred in 770 of Starr's 2239 cases. The statement is commonly made that the left side is somewhat oftener affected than the right, but not all statistics bear this out (Osler, Haven, Sturges, Starr). Another one of the striking features of the motor involvement is the marked loss of power in all muscles affected by chorea. This is especially noticeable in the unilateral and "paralytic" forms, but can be demonstrated also in many of the milder cases, by appropriate tests, such as having the patient grasp one's hand and noting weakness or inequalities of pressure. It is this muscular weakness which explains the fatigue so frequent in choreics.

Involvement of the diaphragm and thoracic muscles results in irregularities in respiration, sighing, etc. which may be graphically represented by means of appropriate apparatus as pointed out by Hauer[49] and later by Graves.[50]* Hiccough, however, is rare. Spasm of the respiratory muscles is also responsible for the peculiar involuntary clucking noises sometimes emitted by choreics. Chorea of the eye muscles is occasionally seen, and when present, may be detected by having the patient fix the vision on some object, while noting the rapid movements of the globe. Very exceptionally, momentary strabismus and diplopia have been reported. Participation of the heart muscle in the choreic twitching is theoretically possible, but at present still lacking of proof. Irregularities in rate may represent an early endocarditis or more commonly merely the effect of the irregular respirations on the cardiac rhythm. Involvement of the smooth muscle fibers in the stomach and intestine, if it occurs at all, is rarely recognized, for increased peristalsis, colicky pains, etc. are very uncommon. Incontinence of urine or feces as a result of chorea is quite unusual even in the severest cases. So also are asthmatic symptoms from participation of the bronchial musculature in the choreic process, even though the other respiratory muscles show distinct involvement.

Skin manifestations in chorea assume the character of those found in rheumatism, the commonest lesions being those of urticaria, erythema multiforme, and purpura. They are infrequent during the entire period of childhood, but especially rare in the younger children. Subcutaneous "rheumatic nodules" are also seen at times with or without other manifestations of rheumatism. Finally, when arsenic is used in the treatment of the disease, it must be remembered that in susceptible individuals this drug is capable of producing a wide variety of cutaneous lesions, the commonest of which is bronzing of the skin. Drug rashes, however, must be composed with clinic manifestations.

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* According to the latter, such irregularities in respiration may persist for many months after all other choreic manifestations have disappeared, and Graves believes their persistence lends support to the view that most of the so-called recurrences are in reality true relapses.

The *knee-jerks* in chorea are quite variable; at times, especially in chorea mollis, they are absent or elicited only with great difficulty, whereas at others, even in the same patient, they may appear to be markedly exaggerated. In the latter instance, an unusually sharp blow on the patellar tendon, or several taps rapidly repeated, frequently causes two or three jerks, or the limb rises to a position of extreme extension, and remains so for a moment or two before becoming flaccid again (the "hung-up" knee-jerk). The knee-jerks return to normal with convalescence. Ankle clonus never occurs even when the knee-jerks are much exaggerated. Other reflexes are normal. Electric stimulation of nerves and muscles is said at times to show an increased excitability. Tactile sensibility is as a rule normal, and anesthetic areas which may be demonstrated in older girls or young adults are usually explainable on the basis of hysteria.

Chorea ordinarily runs an afebrile course, but occasionally moderate degrees of *temperature elevation* may occur, especially in the severer types of the disease. In those cases bordering on acute mania, even hyperpyrexia has been observed. In most instances, however, fever will be found to be due to a complicating endocarditis or arthritis. It must be remembered in this connection, that fever often occurs in acute endocarditis long before the physical signs arouse the suspicion of a cardiac involvement. *Heart murmurs* are frequent in chorea, being usually soft and blowing in character. They may be due to a beginning endocarditis, transient cardiac dilatation, or to anemia, and possibly in some instances to chorea of the heart muscles, although this is still lacking of proof. Only the former is of any serious consequence, however.

The *urine* may show traces of albumin in the severe cases, but this is usually absent in the average case except in the presence of a complicating nephritis or endocarditis. Glycosuria has been reported as a rare complication but is probably merely an accidental finding and A. E. Garrod[51] found urohematoporphyrin in 14 out of 20 cases.

The *cerebrospinal fluid* is often quite normal, but may show a lymphocytosis in some instances. Richardiere, Lemaire and Sourdel[52] claim to have found this present in 12 out of 14 cases examined by lumbar puncture, whereas Morse and Floyd[44] found a lymphocytosis (highest cell count 25 cells) in only three out of 10 cases examined and Quigley[45] in none of 21 cases studied.

The blood in chorea often exhibits signs of a mild or moderate anemia, and in addition may show an increase in the eosinophiles as pointed out by Pianese,[35] Macalister,[53] Leopold,[54] Berger[55] and others.

Complications.—Chief among the complications of chorea, both because of their frequency and serious nature, stand the cardiac affections. The heart is often accelerated and may even be irregular, though free from organic lesions, the disturbance in rate being dependent upon the influence of the irregular respirations on the cardiac rhythm. Functional murmurs, soft and blowing in character, may likewise occur, due to the anemia. Moreover it must be remembered that moderate degrees of transient cardiac dilatation are frequent in the early stages of chorea, just as they are in many other infectious diseases of childhood, and may give rise to murmurs which are falsely interpreted as being due to organic lesions of the valves. Frequently, however, fever, irregularities in pulse rate, and murmurs, develop as a result of true organic disease of the heart, the commonest form being acute endocarditis. This is in every way comparable to the endocarditis of rheumatism, and may be accompanied by pericarditis or myocarditis as well. It is the ever present possibility of one or more of these complications in chorea which makes the outcome doubtful. The frequency with which cardiac lesions occur is difficult to determine, because very mild cases of endocarditis may recover and leave no trace of their former existence, and on the other hand, supposed functional murmurs may persist and prove ultimately to have been due to true organic disease. However, in the series collected by Mackenzie[8] 32 per cent. of the 439 cases had definite cardiac lesions and in addition 16 per cent. had "some derangement of function or change of an indefinite and probably not organic character." Abt and Levinson[11] give the percentage with organic heart lesions as 32 per cent. in 226 cases. Osler[7] reported that of 140 cases examined more than two years after an attack of chorea, 72 or 51.4 per cent. had heart lesions. Our own series showed 23.7 per cent. with definite organic cardiac involvement, and an additional 9 per cent. in whom this diagnosis seemed likely but not altogether proven. In many such cases the electrocardiograph may be of service in showing the extent to which the heart has been damaged.

Tonsillitis and arthritis, whether regarded as complications or merely manifestations of the same disease, are fairly common in chorea. The latter is, fortunately, less frequent than the former, and rarely occurs in cases with severe motor disturbance. Rheumatic nodules occurring during the course of chorea are only infrequently noted in this country, although fairly common in England, but Brennemann[56] believes that at least in other rheumatic affections, they are merely overlooked being almost if not quite as common here as in the British Isles. He believes their presence indicates a severe infection, likely to be complicated by some cardiac involvement. Among the other complications which may occasionally follow chorea may be mentioned nephritis and acute mania, and, very rarely, other forms of insanity.

Diagnosis.—The diagnosis of well-marked chorea is rarely difficult. The irregular, involuntary movements, emotional instability and muscular weakness are so typical as scarcely to be confused with any other condition. Occasionally, however, mistakes in diagnosis may occur, especially when the chorea is very mild or shows an accentuation of certain features which obscure the lesser manifestations. In very mild cases, it may be difficult at times to say whether the movements are choreic or due to the general fidgetiness of a nervous child. Only close observation and the subsequent course of the disease will settle this question. *Habit spasm* or *tic* is sometimes confused with chorea, but in the former the movements are limited to certain groups of muscles are coördinated, purposeful, and consist of repetitions of the same action. Habit spasm runs a much more chronic course, and has neither the varied picture, the psychic disturbances nor the tendency to heart complications seen in chorea. The paralytic form of chorea, especially if limited to one side, may bear a superficial resemblance to *hemiplegia,* but the history of gradual onset, presence of twitching elsewhere in the body, and course of the disease, will usually rule out cerebral hemorrhage or embolism. The movements in so-called post-hemiplegic chorea are as a rule slower, athetoid in character, and there are other signs of a cerebral lesion such as spasticity, exaggerated reflexes, etc. *Poliomyelitis* may be suspected when the paralytic form of chorea affects chiefly one extremity, but the former is more sudden in onset, is usually followed by atrophy of the affected muscles, and has none of the twitching seen elsewhere in the body in chorea. Again, fever, hyperesthesias and changes in the spinal fluid belong much more to the early stages of acute poliomyelitis than to chorea. Chorea mollis has been mistaken for *post-diphtheritic paralysis,* but there are many points of differentiation and with proper observation confusion should not occur. Involvement of the soft palate is an almost constant finding at some time in the course of multiple neuritis following diphtheria, but much rarer and never as marked in chorea. In diphtheritic paralysis, the knee jerks are usually absent or much diminished, but in chorea they are more apt to be normal or exaggerated. The history of onset, course, presence or absence of an unstable emotional state, and choreic movements will aid in establishing the proper diagnosis. *Friedreich's ataxia* frequently presents a coarse tremor and movements which are mistaken for those of chorea. In Friedreich's ataxia, however, there is a marked tendency to hereditary and familial characteristics, very chronic progressive course, peculiar foot deformity and nystagmus, which are lacking in chorea. It affects the lower extremities most, and before the upper ones, whereas in chorea the opposite usually occurs. *Huntington's chorea* does not occur in childhood, shows a marked hereditary or familial character, and runs a very chronic course to pronounced mental deterioration and finally death. *Hysteria* may at times simulate chorea but is much rarer in childhood than in adult life. The movements in hysteria have not the irregular, purposeless character of those in chorea, and are usually accompanied by sensory and other phenomena which will clear up the diagnosis. *Chorea electrica* really bears but a superficial resemblance to true chorea and is not related to it in any way. This name has been applied to a variety of affections characterized by lightning-like contractions of one or more muscles, the effect resembling that produced by momentary electrical stimulation of these muscles.

Course, Relapses and Recurrences.—The duration of the disease is very variable, and its course is usually marked by periods of exacerbation and remission of the characteristic movements. The average duration is from 6 to 10 weeks but not infrequently recovery is not complete for three or four months. In a few instances, the disease continues for a period of a year or even longer, with remissions and relapses. Rarely, its course is completed in two weeks or less. The average case requires one or two weeks to reach its height, and after remaining at about the same level for several weeks, slowly declines again. Often a certain amount of residual chorea remains for several months, or in the most chronic form, for several years. In certain instances choreiform movements of the head and neck may become perpetuated as a *habit spasm.* Although the latter may be more or less permanent, by far the greater number of uncomplicated cases of chorea end in complete recovery.

Recurrences in chorea are very common and are always to be expected. They occurred in over 15 per cent. of Abt and Levinson's[11] 226 cases, in 21 per cent. of ours (220 cases), and in 41 per cent. in Osler's[7] series (410 cases). Osler states further that 60 of these (14.6 per cent.) had more than two attacks. With respect to repeated recurrences, they are by no means rare, Sachs[57] for example, reporting seven patients with four attacks, three with five and one with eight attacks. Cases are on record with 12 or even more attacks, but possibly some of these are relapses rather than recurrences. The disease is most apt to return in the springtime, and it is at this time of the year that children who have had one attack must be watched most closely. Relapses are less frequent than recurrences, but may occur at any time during convalescence.

Prognosis.—In the vast majority of cases, the prognosis of uncomplicated chorea, no matter how violent the movements, is good. Occasionally death takes place in severe chorea, apparently as a result of exhaustion. In those cases with cardiac murmurs, however, the prognosis as to complete recovery must be very guarded, for a large proportion of such murmurs are due to true organic lesions. Even including those cases with endocarditis or pericarditis, the death-rate during acute chorea is very low, probably not over 2 or 3 per cent. The ultimate fate of children with cardiac involvement will depend upon the nature and extent of the cardiac lesion. In rare instances, a fatal outcome has been recorded as a result of hyperpyrexia and septicemia, but it is possible that some of these were in reality cases of unrecognized endocarditis.

Treatment.—**Prophylaxis.**—Although it seems quite likely that chorea is an infectious disease the exact causation of which is dependent upon many factors not yet fully understood, nevertheless it seems probable that attacks may often be averted by the observance of a few simple rules of hygiene and common sense. The development of choreic symptoms should be watched for especially in those children who have had a previous attack of rheumatism or chorea, or in whom there is a strong family history of either of these diseases. In such children, or in thoses with marked neuropathic tendencies, every effort should be made to build up the general health and improve the mental poise. The first of these objectives can often be attained by a proper regulation of home and school life, and intelligent supervision of the diet. Over-pressure in school must be avoided, and plenty of opportunity afforded for exercise in the open air. If necessary the child should be taken out of school for a time and sent to the country. A proper amount of sleep is of the greatest importance, and this should include not only going to bed at a reasonable hour, but also a nap in the middle of the day. In trying to provide a period of deep, restful sleep at night, a point so well known as to be frequently overlooked is the one that the windows be kept open while providing sufficient means for comfortable body warmth. Medicine will rarely be needed, except the occasional use of iron for anemia, some bitter tonic to stimulate the appetite, or cod-liver oil as a general food tonic. The improvement of the mental attitude usually follows as a direct result of the above measures, but at times it may seem absolutely necessary to provide a change of environment as well. The good results will often be in direct proportion to the completeness of this change, and consequently parents and indulgent relatives are rarely the best care-takers under such conditions. All undue excitement, whether produced by school work, books, moving pictures or what not, must be avoided, and tea, coffee, or other stimulants forbidden. Play or exercise in the open air is very desirable, but must be so supervised as to prevent over-fatigue or such exposure as might provoke an attack of rheumatism in a susceptible child. By the observance of such simple rules as these, it is often surprising how much can be accomplished in the way of building up a child both mentally and physically, and while we may never have the satisfaction of knowing that we have actually prevented an attack of chorea, the parents are usually quite as grateful for the general improvement in the child as though the other malady had in reality already existed.

Finally, under prophylaxis must be considered the question of tonsillectomy. Because of the frequent association of recurrent tonsillitis with rheumatism and chorea, plausibility is lent to the theory that the portal of entry for these infections lies in the tonsil. Whether or not this be true, the custom is growing among pediatricians to advise tonsillectomy in those children who have had one attack of rheumatism or chorea, as a possible means of preventing recurrences and lessening the danger of the development of heart complications.

It is difficult to get reliable data on the value of this procedure, but according to St. Lawrence[58] of 40 children in whom tonsillectomy was performed subsequent to an attack of chorea, 20, or 50 per cent., suffered a later recurrence of the latter disease. Too much should not be promised, therefore, as there are times when tonsillectomy is apparently without effect in the prevention of chorea and rheumatism, or even sore throats.

In the treatment of chorea itself, drug therapy is second in importance to measures directed toward the general hygiene. This will include much of what has been said under "Prophylaxis," with especial emphasis on the urgent need of mental and physical rest. In fact rest in bed is the most valuable single remedy we have in the treatment of chorea, and this measure should be employed during the acute stage in all but the mildest cases. Even when the disease is very mild, rest is of great benefit, although it may suffice to take the child out of school and limit the activities to such mild amusements and gentle exercise in the open air as will not overtax the easily fatigued muscles and high strung emotions. It is often an advantage, and in the severe cases usually quite necessary, to have the patient in a room by himself or at least screened off from the other patients; this not only prevents undesirable destractions for the patient, but in a hospital shields the other patients from an unpleasant sight. If the patient is at home, it will usually be desirable to exclude from the room all relatives and friends, except the care-taker. The attendant in charge should, if possible, be a tactful person of placid disposition, capable of exercising a certain amount of moral restraint on the patient without showing undue sympathy. In fact, most chorea patients do very much better in a hospital than at home, and this should always be advised for the severe cases.

When the movements are very violent, the patient must be prevented from injuring himself by means of forcible restraint or padding the sides of the bed. In such instances the patient often experiences considerable relief when the frenzied movements are restrained by pinioning the arms to the sides with a blanket wrapped firmly about the body. The effectiveness of this procedure may often be increased if the blanket is wrung out of hot water first and a dry blanket wrapped outside this. The child may be left in this hot pack for a period of an hour or more provided a careful watch is kept on the pulse and general reaction. It is rarely wise to apply the pack more than once daily, and occasionally a cold pack, applied in much the same way, will be found even more effective than the hot. More recently continuous warm baths, prolonged over a period of several hours, have come into favor. Should these measures seem a trifle too heroic for the milder cases, a prolonged warm bath at bed time will often be found effective in promoting sleep, and gentle massage with a daily cold douche frequently diminishes the movements during the day time.

The question of how long to keep a choreic child in bed is one not easily answered. In general, we may say that it is wise to keep the patient in bed until there is very definite and marked improvement, but not necessarily absence of all choreic movements. In uncomplicated cases this will usually mean a period of from two to four weeks.

Children who still show so much jactitation that they are unable to feed themselves, drink a glass of water alone, or who still exhibit marked weakness of one extremity or one side of the body, should by all means remain in bed a while longer. If, however, the disease has subsided gradually for several weeks and then becomes stationary, with a certain amount of residual chorea still present, the child should be allowed to get up, at first for only an hour or two daily, and the effects of mild exercise ascertained. In certain instances, however, complete bed rest is so irksome to the child that there is danger of the mental excitement converting a mild chorea into a severe one, and in these cases a compromise may sometimes be affected on the basis of rest in bed for part of the day and gentle play for the remainder. During the rest period, the child may be kept amused by means of reading or playing simple games which make no great demands on coördination; but sewing, embroidering etc., which require considerable precision of movements had better not be attempted because of the distressing mental effect when realization of the inability to perform accustomed tasks occurs. In trying to determine how long to keep the patient in bed, the faintest suspicion of cardiac involvement will, of course, prolong this period indefinitely, and bed rest must be insisted upon in even the mildest cases on the first development of a heart murmur, until its character can be studied.

Another point in the general care of chorea which is second in importance only to the imperative need for rest, is that which relates to the state of nutrition. If the disease is at all severe, the patient will usually show considerable wasting, and an earnest effort should be made to prevent this by a careful supervision of the diet. The diet should be somewhat lighter than in health, but of nearly equal caloric value, since the amount of energy expended in muscular movements is sometimes enormous. Great patience on the part of the nurse is required to secure the ingestion of a sufficient amount of food in those patients who have difficulty in swallowing, and it may even be necessary to resort at times to gavage or feeding through a nasal tube.

The **drug therapy** of chorea is disappointing. It may be said at once that there is no specific for chorea, although from time to time arsenic and the salicylates have been acclaimed as such. Because of the close association of chorea with rheumatism, the salicylates are given preference by many, particulary when there is a complicating arthritis or acute endocarditis. Although this class of drugs may be of service in relieving joint pains and perhaps, in full dosage reducing the temperature slightly, the effect on the chorea is, in our experience, usually negligible. If this treatment is to be used, sodium salicylate, or one of the other salicylic acid preparations, must be given in full dose, sufficiently large in most cases to produce tinnitus and slight temporary deafness. This will require for a child of about 10 years initial doses of 5 to 10 gr. of sodium salicylate every four hours, although much larger doses have been advocated by some clinicians. A favorite method of prescribing it is in combination with the bicarbonate of soda, both well diluted. Because of the tendency of large doses of salicylates to cause gastric disturbances, we have tried massive doses per rectum, administered once daily, but have not been impressed with the results.

Arsenic has been used in the treatment of chorea for many years, and by some clinicians is still considered almost a specific. However, we have been unable to convince ourselves of its value, except perhaps as a general tonic in the later stages of the disease. It is best used in the form of Fowler's solution (Liq. Potassii Arsenitis) well diluted. Two drops are given three times a day after meals, and the dosage increased by one drop every day or two until, to an eight year old child, 8 or 10 drops are given three times daily. The dose is then held at this point for another week or two, unless signs of intolerance develop. The attendant should be told to stop the drug at the first sign of vomiting, diarrhea, gastric pain, puffiness about the eyes, or if albumin appears in the urine. In such cases, the drug should be stopped for two or three days and then resumed at the lower dosage and increased to a point just beneath the limit of tolerance. Because of the danger of arsenical multiple neuritis when the drug is given over a long period of time, it is probably wiser to omit this medication one or two days out of every 10. In addition to arsenical neuritis, occasionally a bronzing of the skin is seen following the prolonged use of arsenic, although both conditions are rare. Arsenic may also be given in the form of arsphenamin intravenously, but except in the presence of syphilis, this method has no advantage over others. In one of our cases of chorea with a positive Wassermann reaction, one injection of salvarsan was followed by a prompt cessation of the symptoms.

Perhaps the most useful group of drugs for the more violent forms of chorea are the sedatives, such as chloral hydrate, the bromids, veronal and hyoscine hydrobromid. They are not only valuable in controlling certain of the muscular and psychic phenomena and producing sleep, but at times appear to shorten the course of the disease as well. The tolerance to chloral and drugs of this class is often remarkably increased in chorea, and they must be given in large dosage to produce an effect. Hyoscine is not as commonly employed as chloral and the bromids, but has been well spoken of by certain English and American writers. Antipyrin is also frequently successful in diminishing the movements. Hypnotics such as veronal and trional not only aid in securing restful sleep, but seem to have a general sedative action, and may be given every four or five hours during the day. In the maniacal forms of chorea, however, morphine hypodermically will be found the best drug for general use, along with the wet pack and other measures mentioned above.

Among the many other remedies proposed, simple lumbar puncture has been recommended, but is of doubtful efficacy. The same may be said of non-specific protein therapy, such as the intravenous injection of typhoid or some other vaccin. And finally, Goodman[59] has reported excellent results following the intraspinal injection of the patient's own serum, the rationale of the procedure depending upon the assumption that there may be immune substances in the circulating blood which do not reach the cerebrospinal liquid. The method consists in withdrawing some of the patient's blood, separating the serum, and then injecting 15 to 20 c.c. into the spinal canal after withdrawing an equal amount of spinal fluid. Reports from other observers are too few and conflicting to judge of the true value of this procedure at present, however.

During the course of convalescence from chorea, it frequently happens that after a period of rather rapid improvement, the disease becomes stationary, and for several weeks or months thereafter the patient continues to exhibit a moderate amount of twitching with little or no tendency to complete cure. It will usually be found advisable in dealing with such residual chorea, to terminate the period of bed rest unless there is a heart complication, and under careful observation to permit some form of mild exercise. Should this result in an exacerbation of the chorea, the child should be put to bed again. Gentle massage will be found most useful at this stage, and may often be advantageously combined with a warm bath followed by a cold douche. Should improvement still be slow, exercises intended to reëstablish coördination, patterned after the method described by Fränkel for tabes dorsalis, may be tried. Only the simplest exercises should be prescribed, such as touching the nose with the tip of the finger, fitting wooden pegs into holes, putting a dot in the center of a circle, drawing squares, etc. Some of the easier plaiting and basket-weaving exercises borrowed from the courses on occupational therapy will be found suitable for children, but it is important that all such efforts be intelligently supervised to stimulate interest and yet prevent mental and physical exhaustion.

In addition to such general measures as have been described above, aiming at the relief of the more prominent manifestations of chorea, special symptoms such as anemia, demand appropriate treatment. In fact iron administered because of the anemia, is often the most useful drug at this period of convalescence, and may be the means of completing the cure of the chorea. Recovery will in some instances be hastened by a complete change of environment, especially a trip to some spot in the country where rest and quiet may be secured with an abundance of fresh air and good food, but avoiding such cold, damp climates as might prove favorable to the further outbreak of other rheumatic manifestations.

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CHAPTER CLXXI

FAMILIAL, HEREDITARY AND CONGENITAL DISEASES OF THE CENTRAL NERVOUS SYSTEM

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INFANTILE FAMILIAL PROGRESSIVE BULBAR PALSY

This is an extremely rare affection. Although Londe (1894) refers to several cases beside his own—Fuzio, Hoffmann, and Remak they all presented differences. In the case of Londe, two children, brothers, were affected; in the case of Fazio, the mother, age 22 years, had typical progressive bulbar atrophy, and her child age four years was the case reported. The cases reported by Remak and by Hoffmann were both isolated cases with no family history of the affection. Cases of infantile progressive bulbar palsy, quite similar in their clinical characteristics but without a family history of the affection, have also been recorded by Moty, Hudovernig and Cotton.

The age of onset in the cases reported has varied between four and twelve years. They are normal at birth although stigmata of degeneration have been noted. Neither alcoholism or syphilis has been noted in the parents of these patients. In the case of Remak the symptoms began shortly after an otitis media. In the cases of Hoffmann and of Cotton there was a history of an attack of diphtheria and in the latter also a deafness; but both authors exclude a diphtheritic palsy on the ground that the diphtheria preceeded the bulbar palsy by a year or more. Both of the cases were rather rapidly progressive and fatal.

In the cases reported by Londe the first symptom noticed was inability to completely close the eyes which examination showed to be due to a palsy of the superior distribution of both seventh nerves. The frontalis did not move, and both eyelids drooped though there was no true ptosis. The lips were weak. There was some fibrillary tremor, but no atrophy of the tongue. Deglutition was normal, and the intelligence was good. The optic nerves were atrophic, vision in the right eye was reduced to counting fingers, but in the left eye it was normal. The facial muscles all reacted to the faradic current. In the more advanced case the above symptoms of superior, bilateral facial palsy had added to them the ordinary signs of labio-glosso-laryngeal palsy. In the case reported by Remak one side of the face was affected first, but the upper distribution of both facial nerves soon afterward. This patient had a marked remission of symptoms for three weeks after which they re-appeared with the development of attacks of dyspnoea and tachycardia. In one of these attacks the patient died. There was no atrophy or fibrillary tremor of the tongue and no change in the electrical reactions. These findings together with the remission and the tachycardia attacks makes the case resemble one of myasthenia gravis. There was no autopsy.

The chief difference between the infantile type and the adult type of progressive bulbar palsy is the involvement of the upper portion of the face in the former. The peculiar appearance of the face, inability to completely close the eyes, etc., may resemble the myopathic facies, but the two conditions may be differentiated by the fact that the myopathy is rarely accompanied by bulbar symptoms and extends further to the trunk, shoulder and arm muscles. Post-diphtheritic palsy usually follows immediately after the diphtheria and does not, as a rule, involve the facial muscles. The Moebius "nuclear atrophy' may also affect the upper distribution of the facial nerve, but it is usually noticed at an earlier age and is not progressive. There has been no necropsy on a *familial* case of infantile progressive bulbar palsy, and the pathology is therefore speculative. It seems possible that it is an abiotrophy and may be closely related to the muscular dystrophies. Apparently the only case of infantile progressive bulbar palsy with necropsy is that reported by Cotton, but his case was not familial though showing the same signs and symptoms. No pathology was found in the nervous system outside of the medulla, but unfortunately the medulla had not been examined at the time he published his report.

The disease is apparently always fatal. Londe divides it into three stages. First is the stage of bilateral facial palsy with few other symptoms. This stage is generally short. The second stage includes the addition of the bulbar symptoms; it may last as long as eight months. The third stage is that of inanition, respiratory difficulties, and various complications.

Treatment is unavailing to stop the progress of the disease.

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INFANTILE PSEUDOBULBAR PALSY

This form of bulbar palsy was first described by Oppenheim, and on a pathologic anatomic basis would be classed rather with the cerebral than the bulbar affections. The presence of the bulbar symptoms is dependent on bilateral cerebral lesions. Since all of the muscles affected in the labio-glosso-laryngeal paralysis receive motor impulses from both sides of the brain, unilateral lesion will not cause these symptoms.

In the cases reported by Oppenheim the lesions consisted of bilateral porencephalon, a microgyria affecting the lower portion of the precental convolutions. Similar conditions may be brought about after birth by bilateral encephalitis in this region or in the region of the anterior limb of the internal capsule on both sides. In infants this is frequently due to congenital syphilis.

There is almost always evidence of the cerebral location of the lesion in this presence of infantile hemiplegia or diplegia, and not infrequently, imbecility, epilepsy, etc. In the 28 cases collected from the literature by Peritz only three showed no motor palsy in the extremities. Fickler also reported three cases without involvement of the extremities but all of them were epileptic imbeciles. Fickler claims that this type is characteristic of congenital syphilis and in this respect differs from some of the cases with infantile hemiplegia or diplegia.

The condition may be present at birth or develop later as the result of encephalitis (Brauer) or other lesion, but its development is usually sudden and there is a tendency to recovery. The eye muscles are usually not affected, and the greatest involvement is noticed in the region of the distribution of the fifth, seventh and twelfth cranial nerves. The mouth usually hangs open and the bite is very weak. The pterygoids may also be paralyzed. The facial is affected more in the lower than in the upper portion. The inability to control the lip gives rise to drooling. The soft palate is usually paralyzed, and fluids may regurgitate through the nose. There is no atrophy, and the electrical reactions are normal. The lip, tongue, palate, pharynx and laryngeal movements are interfered with, and there is more or less dysarthria and dysphagia as a consequence. In two of the cases reported by Oppenheim there was trismus and in another complete aphonia.

These cases can be distinguished from the progressive or true bulbar palsies by the absence of atrophy and fibrillary tremor in the tongue. Changes in the electrical reactions do not occur in the pseudobulbar palsy, and it is non-progressive.

The prognosis is not good for the causal condition cannot generally be reached and treatment is therefore ineffectual.

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MÖBIUS INFANTILE NUCLEAR ATROPHY

This term was first used by Möbius to classify some cases observed by him in which the chief symptom consisted of more or less inability to perform the movements of parts supplied by the cranial nerves. The name was based on the assumption that there was an atrophy, total or in part, of the nuclei of the floor of the aqueduct of Sylvius and fourth ventricle. Instead of a nuclear atrophy in these cases there is more often a nuclear aplasia, as Huebner has pointed out. The same symptoms are also produced by atrophy or aplasia of the nerve and by muscle aplasia. In fact many of these cases, especially the ones in which simply one muscle or one movement are affected, are due to congenital muscle defect. In order to include all of these cases it would be more appropriate to designate them infantile movement defects in the region of the cranial nerves (Vogt, Heuck).

The etiology of these cases is unknown. They are frequently hereditary or familial although often not directly. A history of other type of nervous disorder, epilepsy, etc., is frequently met with. It is not uncommon to find other anatomic defects in the same patient. Alcoholism or syphilis in the ascendents "plays no specific" rôle (Vogt). Cases in which there is a definite exogenous cause for a partial bulbar palsy—acute encephalitis, etc.—are not included in this category.

Perhaps the most frequent symptom is congenital ptosis which may be unilateral or bilateral. It may be present constantly or only at certain times as when fatigued or after excessive use of the eyes, etc. All or any of the motor cranial nerves or almost any part of them may be affected. A great number of combinations have been reported. The sensory nerves are rarely affected although the sensory portion of the fifth is occasionally involved. Cases of congenital color blindness, amblyopia, etc., might possibly also be included here. Their association with the movement defects are not uncommon. An isolated weakness in the external rotation of the eye-ball (Axenfeld, Graefe, Kunn) is frequently found. The internal ocular muscles may also be affected; congenital immobility of the pupil has been reported by Oppenheim and also by others. All kinds of combinations of external and internal ocular palsies can occur and be combined also with congenital nystagmus.

Another frequent condition is the congenital facial palsy. This is peculiar in that the upper portion of the face is much more affected than the lower. According to Bernhardt, this gives a very characteristic appearance to the face with the upper portion mask-like, motionless and often appearing sunken.

A classification of the disturbance of movement in the region supplied by the cranial nerves is given by Vogt:

1. Congenital cases of which the signs are noticed at birth or a short time after. They affect chiefly the eye muscles or the distribution of the seventh cranial nerve or both, but there are cases with sensory symptoms and also cases in which all the bulbar nerves are affected.
2. Disturbance in movement in the same region (cranial nerves) in childhood may be progressive. These cases are usually noticed later and are similar to progressive bulbar palsy in adults.
3. Those cases in which the medulla is affected by exogenous factors, trauma, tumor, encephalitis, etc. In these cases there are signs of the causal condition in addition to the movement defect.

Although in the severe cases with marked bulbar involvement life may be maintained with some difficulty, the affection is not considered fatal. In the slight cases, ptosis, etc., the condition is not especially troublesome though it probably will continue through life.

Treatment is not satisfactory, but in the case of a ptosis or a strabismus, operations may give good cosmetic results.

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FAMILIAL DISEASES OF THE MUSCLES

Under this title are included those cases in which there exists a degeneration of the muscles that is usually progressive and is not due to injury of the peripheral nerve or to an acute neuritis. Cases in which the muscle degeneration is due to spinal cord injury, hematomyelia, acute anterior poliomyelitis, diffuse or transverse myelitis are excluded. It is often the case that this progressive muscular degeneration is part of the clinical symptoms of other conditions such as for instance amyatrophic lateral sclerosis, multiple sclerosis, etc., which may or may not be familial.

Cases of chronic anterior poliomyelitis and chronic bulbar paralysis, in which the first symptom may be a muscular atrophy, are difficult to exclude from the above category and are included by some authors (Dejerine and Thomas, Allen Starr).

Historical.—According to Bruns the first mention of progressive muscular atrophy is made by Bell, but perhaps the first fully described case is that of Duchenne, in 1849. A similar case was described by Aran in 1850. Both of these cases belong to the type which we now speak of as the Aran-Duchenne type of spinal progressive muscular atrophy; although at that time they were regarded as primarily muscular. Duchenne, in 1861, described a case of pseudo-hypertrophic paralysis. Since that time a great variety of forms of muscular atrophy have been described, and the myogenous type has been divided and subdivided, though it is recognized that the distinctions between these types are not always sharp.

Spinal Progressive Muscular Atrophy.—Spinal progressive muscular atrophy begins generally about the twentieth to thirtieth years of life. The etiology of the affection is unknown. Trauma may be the cause, as in the cases reported by Ziehen and by Hoffman. Overuse of the part may also be a cause though most of the cases that are so described are in reality cases of occupation atrophy and are not progressive.

The typical clinical picture of the condition has been reported as due to syphilis but most of these cases are cases of chronic anterior poliomyelitis and the atrophy is consecutive to a flaccid type of paralysis. Cases of progressive muscular atrophy of the spinal type due to syphilis are reported by Dana, Leri, Leopold, Souques Baudouin and Lantueqoul and others, and the diagnosis from the non-syphilitic cases is most difficult unless a good history of the onset of the trouble is obtained. In a case of this kind that I reported the patient stated definitely that she had lost the use of the hand before she noticed any atrophy which is quite the opposite to the course of events in the ordinary progressive muscular atrophy. This patient had no other clinical signs of syphilis except an Argyl-Robertson pupil. The Wassermann reaction in both her blood and spinal fluid was strongly positive and her husband had tabes. In my observations on congenital syphilis I have never encountered a case which presented the clinical picture of the spinal type of progressive muscular atrophy although I have seen a number of cases of flaccid paralysis followed by atrophic changes in children with congenital syphilis. Such cases are often amenable to treatment.

Pathologic studies show that the essential lesion in these cases is a degeneration and gradual disappearance of the anterior horn cells of the spinal cord. There is also usually some degeneration in the anterior nerve roots. Histologic examination of the muscles show a simple atrophy of the muscle fiber with overgrowth of connective tissue.

The atrophy begins in the small muscles of the hand usually first in the opponens pollicis and interossei, is not associated with pain and is not preceded by any special disability. The atrophy is frequently noticed first by accident. As it progresses there is a progressive weakness in the use of the hand, but no complete paralysis until the muscle has disappeared. The flexor muscles are generally the first to be affected and the result, in advanced cases, is the so-called "claw hand" with the first phalange extended and the two end phalanges flexed. In a few cases the extensors have been involved before the flexors (Potts). There is usually a fibrillary tremor in the affected muscles and also in those which will be affected though they may then show no atrophy. The atrophy does not progress up the arm in a regular manner, but jumps from group to group of muscles usually from the hand to the shoulder muscles. The shoulder muscles may atrophy before the hand muscles (type Vulpian-Bernhardt). A case of this kind has recently been described by L. F. Barker under the title "Vulpian-Bernhardt Subvariety of the Aran-Duchenne Type." It is fairly symmetrical, but begins in one hand before the other. The tendon reflexes are diminished or lost. The electrical irritability of the muscle is at first diminished and later may show a reaction of degeneration. There are no sensory changes.

Spiller says that chronic external ophthalmoplegia is a form of progressive muscular atrophy of central origin of which there are many cases in the literature. It usually occurs in infancy, though sometimes later and is called by Marburg the "Ponto mesencephalic form." There is some doubt as to the existence of this condition as an isolated affection. Certainly most of the cases of chronic progressive nuclear ophthalmoplegia are but complications of tabes or cerebrospinal syphilis. Studies of this condition have been made by Siemerling, Cassirer, Abrahamson and others. In the cases reported by Ayres, Fuchs, Gowers, Homen and Nazari-Chiarini the condition was familial.

Differential diagnosis must be made from injury to the peripheral nerve in which case there is usually a history of injury. It must be remembered, however, that some injuries are not noticed by children or that in case of fractures of the bones with callous formation, the nerve may be included in the slow development of the callous. The diagnosis must also be made from lead poisoning in which case there is usually more paralysis, wrist-drop, etc., and there are also other signs of lead intoxication. The diagnosis must also be made from a cervical rib which pressed on the brachial plexus and produces a chronic degenerative neuritis. Spiller and Gittings have reported a case of progressive muscular atrophy of a cervico-bulbar type, occurring with cervical rib, in which there was extensive atrophy in both arms. The other causes of a progressive muscular atrophy of the upper extremities are usually organic diseases of the nervous system which are recognized by their characteristic symptoms.

The atrophy that follows the paralysis of acute anterior poliomyelitis while progressive in the muscle affected does not tend to spread. Potts, however, was able to collect 28 cases in the literature in spinal type of progressive muscular atrophy followed acute anterior poliomyelitis though usually, many years later.

Progressive spinal muscular atrophy generally runs a slow course and is subject to remissions.

Treatment is not very satisfactory. The use of strychnin in ascending doses, to the limit of tolerance, may do some good. Electrical treatment, massage, etc. is usually of no benefit and if carried to excess may be harmful. The patient should carefully avoid overuse of the part.

Familial Spinal Progressive Muscular Atrophy.—This disease is rare. The first case was described by Werdnig in 1891. Similar cases were described by Hoffman in 1893. The latter author in 1897 described 20 cases in one family, but it is rarely definitely hereditary. Only in one family, that described by Hoffman, did the disease appear in more than one generation. A case was described by Werdnig in which two children of the same mother, though of different fathers, had the disease. The children usually die in comparatively early childhood.

The symptoms of the affection are usually quite definite. The child is born normally and appears to be normal. Only in the case described by Beevor was there some weakness at birth. The symptoms usually begin in the second half of the first year just after or before the child has begun to stand or walk. It may be first noticed that there is a stamping gait, but the child looks well and there is no fever. From this on the symptoms usually progress fairly rapidly. It differs from the spinal muscular atrophy of adults in affecting more the trunk muscles than the extremities. There develops gradually a difficulty in holding up the head or in sitting up. Occasionally the bulbar muscles are affected (Bruns, and Werdnig). The trouble is usually symmetrical. The weakness of the muscles goes hand in hand with the atrophy and with a certain amount of myatonia. There is usually a fair amount of fat though in the later stages the subcutaneous fat may also disappear. Fibrillary tremors have been observed, but are rare. Electrical reactions of the muscles are changed. There is generally a slow galvanic, and absent faradic reaction in the affected muscles (Bruns). The tendon reflexes are diminished or lost. There are no sensory changes. The sphincters are not affected and intelligence is not disturbed. There is no special tendency to deformity except that from the position assumed by the patient as a result of his weakness. From long sitting in one position a most intense scoliosis may develop.

The course of the disease is progressive and usually death occurs by the fifth year, although cases have been described of longer duration. Death is generally due to some lung affection, the development of such conditions being encouraged by the weakness of the respiratory muscles.

Anatomic investigations have usually shown a degeneration of the anterior horn cells of the spinal cord. The blood-vessels are not affected. The muscle fibers show a simple atrophy with some fatty degeneration. Beevor reported a few hypertrophic fibers mixed with the atrophic.

The diagnosis of the infantile spinal progressive muscular atrophy is not difficult when taken in connection with the familial character of the trouble. In the hereditary neural muscular atrophy the disease does not begin so early in life; it is much slower in development and affects the lower extremities and does not affect the body until later. The pseudo-hypertrophic type of myopathy is not so rapid in its progress; it does not usually begin before the third or fourth year; and there is distinct hypertrophy. Some of these cases might be mistaken for cases of anterior poliomyelitis, but this affection begins acutely, with fever, and the paralysis comes on suddenly and has a tendency to recovery rather than to be progressive. It is very rare to have multiple neuritis in children of this age. On account of the atonicity of the muscles and possibility of scoliosis there is some difficulty in distinguishing it from rickets. Myatonia Congenita (Oppenheim) resembles the Werdnig-Hoffman type, but in the latter the affection is not present at birth and in the former there is a frequent tendency to improvement. Beevor suggested the possibility that a traumatic hematomyelia at birth might produce similar symptoms, but in such cases the symptoms would not be progressive. The condition can be diagnosed from amaurotic family idiocy by the absence of blindness or mental changes.

No treatment that has so far been suggested has appeared to be of any benefit.

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Neural Progressive Muscular Atrophy (Charcot-Marie-Tooth. Peroneal Type).—Cases of this form of progressive muscular atrophy were first described by Eulenberg in 1856. Bamberger in 1860, Friedreich in 1873, and in the same year Eichorst reported a similar case in which this affection had lasted through six generations. In 1889 Hoffman collected the cases already described and reported a case with autopsy, finding the site of primary process in the peripheral nerves. In 1891 Hoffman reported a case which began in the hand.

Neural progressive atrophy is a familial and hereditary disease. Herrington reported 26 cases in one family with numerous examples of direct heredity. In other cases the parents may be normal and the disease skip a generation. There are also a number of single cases reported. In some families only the males are affected. I have observed a family of this kind in which the males were affected with this form of atrophy and the females with pernicious anemia. Occasionally the disease appears to begin after infectious disease, especially measles. A case is reported by Francais in which the affection began in a soldier 10 months after he was injured in both feet by shell fragments. His injury was followed by long suppuration and there was no family history of this affection. The case was not typical. I have seen a case begin shortly after a "stone bruise" on the heel, but regard it as a mere coincidence.

It has been reported as occurring in the negro (Bassoe and Ryerson), though it is not common (Eschner).

It usually begins about the fourth or fifth year, but cases have been described as late as the thirtieth or fortieth year. In the same family the individual cases usually begin at about the same age. The disease, in general, is slowly progressive, but subject to long periods of remission. Occasional cases are rapidly progressive.

The first symptom is usually a slight difficulty in walking due to weakness in the foot muscles. It is generally symmetrical. The peroneal muscles, and the extensors of the toes are first affected, the calf muscles not until much later. When the hands are involved it is usually later in the disease although cases have been reported by Hoffman, Dubreuilh and others in which the hands were affected first. In many cases the hands are never affected. The quadriceps femoris muscle is frequently affected (Sachs, G. W. Jacoby, Given Campbell, Diller) and it may be noticed that the lower zone near the knee is much more affected than the upper zone. Hypertrophy of the muscles does not occur and there is usually no marked development of fat. Fibrillary tremors have been described by some authors. The myotatic irritability of the muscles is usually diminished and sometimes absent. The Achilles tendon reflexes are usually absent. The knee reflex is retained until the quadriceps is affected, but it also disappears in the later stages.

The electrical examination of the affected muscles shows the reaction of degeneration. The faradic irritability of the nerve is also diminished. It has been noticed that electrical changes in the muscles may precede any apparent atrophy.

Lancinating pain is occasionally complained of and also hyperesthesia. Objective sensory changes are rare, but have been described. The skin reflexes usually show nothing abnormal. The nerves are not tender on pressure. The feet are frequently cold and appear cyanotic. Sweat secretion is often increased. The sphincter and mental functions remain intact. There is generally a "steppage gait," the knee is overflexed and the foot raised so that the patient appears to be stepping over something. The toes strike the floor first, but are not dragged or scraped and the heel comes down with a thump." The movements of the body and head are usually not affected and the point is made by Charcot and Marie that in this type the muscles near the trunk are relatively little involved and that the shoulders and face always escape. The feet generally have the deformity of pes equinovaris, a deformity resembling somewhat the congenital club foot. In a few cases there has been observed a fine tremor of the hands. Charcot and Hoffman noticed a tremor of the face and tongue. The disease may be combined with optic atrophy and also with fixed pupils (Oppenheim). Cases have been described as occurring in combination with Friedreich's ataxia and other heredo-degenerative conditions.

Hoffman on the basis of two cases with necropsy described the pathologic anatomy as degeneration in peripheral nerves. In both cases he found also a degeneration of Goll's tract in the spinal cord. The muscle fibers showed a fatty and nuclear degeneration also an absence of muscle fibrillæ. Similar findings have been reported by Dubreuilh and others. In cases which clinically resembled a neural progressive muscular atrophy, Oppenheim and Cassirer found no changes in the nerves or the spinal cord, but only changes in the muscles. The pathology of the condition can not be regarded as settled, but probably Oppenheim's case was a myopathy of the distal type (which see).

The diagnosis of neural progressive muscular atrophy is generally not difficult. When it begins in the hand it may be mistaken for the spinal type, but the ordinary spinal type is not usually familial and does not begin at such an early age, whereas the infantile spinal type which has been described above is much more rapid in its progress and begins usually in the body. It is much more difficult to diagnose this condition from chronic multiple neuritis, especially lead neuritis which in children usually affects the lower extremities. Lead neuritis is not familial nor hereditary and would usually present other symptoms of lead palsy. The history will exclude club foot. It is necessary to diagnose from this condition the disease reported by Dejerine and Sottas as *hypertrophic progressive interstitial neuritis of infancy.* This disease begins in childhood with atrophy and paralysis of the muscles of the distal end of the extremities. Also, there are deformities of the feet. The tendon reflexes are lost. There are severe lancinating pains, distinct sensory symptoms, ataxia and Romberg symptom. A characteristic of the disease is the distinct thickening of the nerves. Anatomically there is marked increase in the interstitial connective tissues of the nerves accompanying their degeneration. This disease is also occasionally familial. It is, both clinically and anatomically, closely related to the neural muscular atrophy.

The duration of the neural muscular atrophy is indefinite. The progress is extremely slow. It does not appear to shorten life and may become permanently halted. Treatment is not of much avail. The use of electricity and intramuscular injections of strychnin may be of some benefit. In event of great deformity, orthodesis or tendon transplantations may be necessary.

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PRIMARY MYOPATHIES

The primary myopathies differ from the spinal progressive muscular atrophy and from the neural type in not showing evidence of changes in the nervous system either anatomic nor functional. They were among the first of the muscular atrophies to be described though, at that time, they were not divided from the spinal type. A number of different varieties of myopathies have been described but differing from each other only as to the localization of the changes. The nature of the process is probably the same in all cases and the recent tendency has been to consider them as one general disease. It is probably better, however, to retain the classification into different types if only for the reason that it facilitates their description and recognition.

Etiology.—From a study of the cases reported it is quite evident that the chief factor in the cause of the trouble is a congenital tendency. For the most part they are distinctly hereditary or familial although they do not conform to the "Mendalian laws" of heredity. It would seem more likely that the process really consists of a congenital tendency to degeneration due to a fundamental defect in the germ plasm for which Gowers has used the term "abiotrophy." There is also an evident relation between the myopathies and familial degenerative diseases of the nervous system. I have reported a case of a patient with progressive muscular atrophy who had descendants with a myopathy and similar cases have been reported by Hertz and Johnson and by others. I have also observed a case of progressive pseudohypertrophic dystrophy combined with Friedreich's ataxia and a case of myopathy combined with hereditary optic atrophy. Various other combinations of similar character have been reported in the literature. The cause of this congenital tendency is unknown. It would seem likely that the real cause lies in a blastophoria which originated in some toxic or infectious process. In such a case (for instance lead poisoning) the off-spring in the first generation show the effects by small size, a general weakness and lower resistance to infections. The next generation not infrequently shows anatomical and degenerative changes. Among the toxic factors which should be considered would be alcohol, lead, etc., but studies along these lines have not been sufficient to incriminate any special one. Attempts have also been made to show that hereditary syphilis may play a part. Leopold found evidence of hereditary syphilis in six of 29 cases of dystrophy, but he does not show that there is an etiologic relation between the two. Cadwalader and White found only one case in seven gave a positive Wassermann reaction and in the others there was no evidence of syphilis. General nutritional disturbance in the parents would not seem to be sufficient to act as a cause and the chemical studies which have been made of the metabolism in myopathies have not cast light on the etiology. Attempts have been made by Timme and others, to show a relation between the endocrine glands and the myopathies but without much success. The symptoms of a myopathy are so distinctly different from any of the recognized syndromes due to change in internal secretion and the mode of development is so different that any causal relation can be practically excluded. Navarre and Correas could find no pathologic changes in the endocrine glands from cases of myopathy. In a case of juvenile type reported by J. Mitchell Clark there was enlargement of the parotid gland and all the salivary glands were enlarged in cases reported by Schönborn and Sacara-Tulbure.

Guillian believes that typhoid fever may be a cause of myopathy. Some of these cases (Friedlander, Lesage, Cerne, Babinski) may be an angio-myopathy due to phlebitis but others are progressive. In this latter type Guillian includes only the cases reported by Friedlander, Josserand and his own. Guillian concludes that myopathies may be acquired, possibly from infections or intoxications.

The myopathies are not especially rare. It is likely that mild cases are frequently overlooked and that the number of reported cases does not by any means represent the total occurrence of the condition. They usually develop at an early age although they may occur even as late as 40. Males are more frequently affected than females and the onset is slow and insidious.

Pseudohypertrophic Type.—In the description of the types of myopathies the pseudohypertrophic type takes the first place because of its frequency and because it was the first to be described, by Duchenne. This type is usually familial, sometimes directly hereditary. The symptoms usually begin about the age of five or six years, but the onset may be delayed until much later. Usually the first symptom noticed is a difficulty in gait. The child does not run with the former facility. He frequently loses his balance and falls and when he walks there is a tilting of the pelvis giving a waddling appearance. It may also be noticed that when he falls he has considerable difficulty in getting up. As the condition progresses the manner of rising becomes characteristic. The child lying flat on the back first rolls over on the stomach and in advanced cases this may be difficult and only possible with assistance. He then rises on the elbows and then on the hands, then on the hands and knees. From this position he gets to a position in which he stands on his feet, but with his hands still on the floor. With considerable effort he gets both hands on his ankles and then pushes his body up with his hands against his legs, gradually assuming the erect position. It is quite evident that this manner of rising is due to weakness in the erector spinæ and gluteal muscles. When standing there is a characteristic lordosis of the spine and largely because of the lordosis, a prominent abdomen. The feet are usually planted wide apart and it will be noticed that the calf muscles are disproportionately large in comparison with the thigh muscles or other portions of the body. The calf muscles may be felt to be hard and firm. The pseudohypertrophy may also occur in other muscles especially in the infraspinatii, deltoids, triceps and glutei.

The condition usually progresses to a weakness of the shoulder muscles which causes an abnormal position of the shoulder giving rise to the so-called loose shoulder. The point of the shoulder is dropped and when the arms are extended forward the scapulæ project backward, like wings. If the child is picked up with the hands under the axilla it will be noticed that the shoulders seem almost to leave the body. The weakness of the shoulder is chiefly due involvement of the triceps, pectoralis major, latissimus dorsi and serratus muscles.

A characteristic symptom lies in the atrophy of the muscles. This atrophy is usually sufficient to be noticed readily, but may be covered by the deposit of fat. It is quite characteristic to find atrophy and pseudohypertrophy in the same muscles and this is especially true of the deltoid.

The atrophy generally affects the triceps, except the upper portion, the pectoralis major, the serratus, latissimus dorsi, biceps quadriceps femoris and adductors. Occasionally the atrophy affects the muscles longitudinally, as well as transversely, and the consequent shortening may give rise to some deformity (Oppenheim).

When the face is affected there is usually a depression in the muscles under the cheek bone. The face becomes expressionless. The obicularis oris is relatively little affected so that the lips project, giving the so-called "tapir mouth." It may be impossible to completely close the eyes. The general expression is quite characteristic and has received the name "facies myopathica." There have been cases in which bulbar paralysis has occurred (J. Hoffman), but these cases are perhaps cases similar to those formerly mentioned in which the usual type of myopathy is combined with some other hereditary degeneration. Oppenheim has described cases in which the tongue, palate, laryngeal muscles, etc., have been affected. The eye muscles are very rarely affected though cases have been observed by Gowers, Oppenheim, Marie and others. Cases have also been reported (Ross, Hammond and others) in which the heart has shown hypertrophy, but Oppenheim doubts this condition. Haushalter has reported two cases with autopsy in which the heart showed degenerative patches in the muscle tissue. In view of the frequency with which the rate and rythm of the heart beat is affected in the later states it is strange that anatomic changes are not more frequently reported. Bunting seems to have been the only other observer to report such a finding. The affection involves both sides of the body but it is not always symmetrical.

The reaction of the muscles to the electrical current is usually normal. There may be diminution of the irritability to faradic current, but never a reaction of degeneration. The tendon reflexes are preserved until the later stages when the muscles are much atrophied. Sensation is normal, likewise the function of the sphincters. Fibrillary tremors are absent.

A number of cases have been described in which there have been congenital anomalies in various parts of the body affecting bones or other parts. Cases combined with atrophy of the bones have been reported by Friedreich, Schultz, Lloyd, Spiller and others. In 1911 Hutinel described a condition of "osteo-muscular dystrophy" as a separate type. Its clinical importance is due to its resemblance to rickets or to osteomalacia. Tixier and Roederer also reported three children with the same disease in one family.

The mental state of these patients is usually normal at least superficially, although recent studies (Schukowski) have shown that in some cases there is a certain degree of mental retardation which, however, is not parallel with the muscle involvement.

Juvenile Type (Erb).—The juvenile type begins generally in youth or early middle age, not quite so young as the pseudohypertrophic type. The chief point of distinction is the early involvement of the shoulder girdle and upper arm.

Facioscapulohumeral Type (Landouzy=Dejerine).—This type is characterized by the early involvement of the facial muscles. This usually is soon followed by the involvement of the shoulder girdle and arm. This type was called the "infantile type" by Duchenne, who first described it in 1883, a term which was also applied to the pseudohypertrophic type.

Hereditary Type (Leyden=Möbius).—This type is characterized by the marked hereditary tendency and usually develops in the eighth to twelfth year. The weakness is usually first noticed in the pelvic region, as in the hypertrophic type, but the hypertrophy is absent.

Zimmerlin Type.—The thoracic muscles are first affected and the lower extremities show no changes. It begins from the eighteenth to the twenty-fifth year and is distinctly familial. In Zimmerlin's cases four were in the same family.

Distal Type (Gowers, Spiller).—This type is rare though it is generally accepted as a myopathy. Cases have been reported by Gowers, Spiller (two), Dejerine and Thomas, Oppenheim and Cassirer, Batten, Cotter and Naville. The chief difficulty lies in distinguishing it from the neural or peroneal type of progressive muscular atrophy which it closely resembles. Spiller emphasizes the point that in the neural type the atrophy is sharply limited to the parts below the elbows and knees while in the myopathy the atrophy sooner or later extends to the thighs and shoulder. Batten makes the points that there are no sensory disturbances in the myopathies as there are in the neural type and that the latter never affects the face. The case described by Batten began at the age of three years. One case of Spiller's began at nine years, the other not until 26. Gower's case began at the age of 10 years. In Spiller's case the knee jerks were lost and there was no plantar reflex. A familial tendency is apparently not marked. Spiller found at necropsy that the brain and spinal cord showed no pathologic change. In the muscle there was no recent degeneration, as shown by the Marshi Method, but the atrophy of the muscle fibers was pronounced. There was an increase of connective tissue and fat. The muscle fibers that remained preserved their transverse and longitudinal striations and the nerves in the section were normal.

Myotonia Atrophica.—A rare disease or type of myopathy in which the myopathy is combined with a myotonic condition in some or all muscles. Hoffman was able to find about 80 cases reported in the literature up to 1912. The disease has been studied by Batten and Gelb, Ascenzi, Hoffman, Curschman, Greenfield, Klineberger, Kennedy, Spiller and others. The pathologic changes in the muscles resemble those of myopathy. It generally affects members of a family in the same generation and is more frequent in males. The myopathy often resembles the distal type. Hoffman found that 10 per cent. of these cases had congenital cataract whereas hereditary cataract is not seen in association with true Thompson's disease (myotonia congenita).

Hypertrophia Musculorum Vera (Dystrophia Muscularis Hyperplastica.—This condition usually affects young adults but the case reported by Brucks was in a child 20 months old. A general increase of bulk of the muscles is associated with weakness. In the case reported by Woods the histologic examination showed a hypertrophy of the muscle fibers but no degeneration. The electric reactions are normal.

Differential Diagnosis.—In marked cases which are typical and uncomplicated the diagnosis is easy, however, it must be remembered that the muscular atrophy is sometimes covered by fat in infants or young children. The chief difficulty in diagnosing comes in differentiating between the myopathies and spinal progressive muscular atrophy. In some cases it is extremely difficult to classify these cases because of the presence of some signs of other conditions.

As before stated it is very probable that the morbid process is very much the same in all these cases. It is characteristic of the myopathy, however, to be hereditary, to show no fibrillary tremor, to begin in youth or childhood and to show the greatest affection in the muscles attached to the trunk, whereas the muscles of the periphery of the extremities are affected later if at all.

A localized or diffuse muscular atrophy may occur as the result of polymyositis. The history in these cases, however, pain, tenderness, etc., and the fact that they are not generally progressive will enable the diagnosis to be made. The diagnosis may be difficult in young infants, but the peculiar way in which the body is held in walking is often diagnostic. A somewhat similar position may be assumed as the result of inflammatory disease of the spine or any painful affection in the back. An acute poliomyelitis may affect the back muscles and give rise to a similar peculiar gait. Osteomalacia and rickets must also be taken into consideration in the diagnosis. Chartier has reported a case of periscapular muscular atrophy which resembled an early stage of myopathy but which was due to beginning pulmonary tuberculosis.

A congenital muscular defect may also give rise to some difficulty in diagnosis though they are generally localized and not progressive.

Progressive muscular atrophy may be combined with imbecility or epilepsy. Oppenheim has reported a combination with tabes and Cassirer a case combined with poliomyelitis. It is probable that these cases are coincidences.

Prognosis.—The prognosis as for life is usually good. It is only when the respiratory muscles are so much affected that there is marked dyspnea that there is danger of a fatal lung complication. The condition is slow in its course and may not shorten life especially if it does not begin too early. Oppenheim reports observing cases in patients as old as 48 years who have had the condition almost all their lives. The disease may be arrested at any point in the course. Abortive forms may occur in which only a few of the symptoms are present. In a few cases improvement has been said to occur. Erb reported a case of the juvenile type which he said was "practically cured" in six months. Jendrassik reported two cases with recovery and states that in his cases as in the cases reported by Marina and by Armand-Delille the improvement occurred at the period of greatest physical development, i.e., puberty. Spiller reports two cases in which there was marked improvement. In one of them there was a positive Wassermann reaction on the blood. The atrophy was of the Erb's juvenile type and the improvement was apparently due to massage and physical therapy, no anti-syphilitic treatment being given.

Treatment.—Treatment is generally not very satisfactory. Excessive muscular strain must be forbidden, but a certain amount of exercise is generally considered beneficial. General attention to hygiene is always desirable, especially good feeding. Oppenheim reports that he has seen a decided improvement result from the use of galvanic electricity and gymnastic exercises, but Marie warns against the use of electricity. Rossilimo has suggested treatment with thyroid tablets and has noticed improvement. Marinesco found no result from the use of thymus. There is usually no deformity and orthopedic surgery is usually unnecessary. Eiselberg has noticed improvement in the use of the arm by performing an operation to fix the shoulder blade but orthopedic apparatus would probably do just as well.

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MYOTONIA CONGENITA (THOMSEN'S DISEASE)

Thomsen's disease is an affection which is almost always hereditary, although a few isolated cases are on record. In the family of Doctor Thomsen, who first described this condition, there were 20 cases in four generations. The condition is usually first noticed in early childhood, sometimes not until as late as puberty or even later (myotonia acquisita). It is probable that in all cases it is congenital, but that in the cases noticed late the condition has remained latent. In some of these cases its development has been due to psychic shock.

Symptoms.—The chief symptom is the difficulty in movement after rest. The first attempt to move the muscle will result in an inhibition of the voluntary act so that the muscle remains rigid. The tonic state may be present in only one group of muscles or in all. The condition may be so marked that the attempt to raise from the chair produces a general stiffening of the body and the patient falls to the floor. Usually in about 3 to 10 seconds the hypertonia passes away and the part can be moved without difficulty. The patient can then walk along distance, dance, etc. Whenever the patient attempts suddenly to make a quick movement the hypertonia returns. In cases where the affection is slight the patient can avoid all difficulty in timing all movements at a very slow rate. The face and palate muscles are rarely affected although it is possible that some cases of stuttering might fall in this category. In a case reported by Charcot the eye muscles were affected. Oppenheim states it is doubtful if the heart is ever involved. The respiration is rarely involved. The hypertonia is increased by excitement and also by being quiet. Oppenheim also found it increased in fevers and especially in cold places. In one case which he reports it was present only in the cold. The objective examination of the patient shows an apparently normal musculature, usually very well developed. The muscular irritability is generally much increased while the irritability of the nerve is not changed. Percussion of the muscles with a percussion hammer gives rise to a tonic contraction of the part struck, which is of considerable duration. The faradic irritability of the nerve is generally not altered. The direct faradic irritability of the muscle is increased and a weak current produces a contracture of long duration. The direct galvanic muscular irritability is also increased and has a somewhat tonic character. A constant current often produces a rhythmic undulation of the muscles.

The above described symptoms may be practically the entire clinical picture and the other functions of the nervous system not influenced. When the eye muscles are affected, von Graefe's phenomenon may seem to be present. A number of investigations have been made with reference to chemical changes occurring in the blood and urine in myotonia congenita. Although an increased amount of creatinin has been found by some, this has not been confirmed by others. The condition is not uncommonly complicated by mental changes, epilepsy, migraine, etc., also with pseudohypertrophy and other forms of muscular dystrophy (see myotonia atrophica).

The **diagnosis** is generally not difficult. The myotonic electrical phenomenon is characteristic. Eulenberg has described a condition which he calls "paramyotonia congenita." This condition is characterized by stiffness of the face and neck and extreme muscular weakness of the obicularis oris and obicularis palpebarum whenever the patient became cold. No increased mechanical irritability of the muscle was observed. Somewhat similar cases have been described by Martius under the title of "myotonia congenita intermittens." Talma described a condition as "myotonia acquisita." The patient would only develop myotonic symptoms after energetic exertion. Cases in which the myotonic electric reaction was absent have been reported by Fürstner and Bechterew. Cases have also been described in which no increase in mechanical irritability of the muscle has been shown (Seligmuller, Strumpell and others). The relation between myoclonia and myotonia has been described by Lundberg. Oddo has described cases of myotonia with intermittent paralysis. A large number of cases somewhat resembling myotonia but difficult to classify have been reported. It is quite evident that congenital myotonia is an affection with numerous variations and extensive relations.

The **prognosis** for life is good. It is not at all certain that these patients can recover although a few cases are reported in which there have been marked remissions. There is generally no progressive tendency. The pathologic changes that have been described were entirely in the muscles. Schiefferdecker gave the following description: "The muscle fibers are larger than normal although among the hypertrophied there may be many small fibers. The increase of nuclei is only so great as the increase in the size of the fiber so that the relative proportion is unchanged. The sarcoplasma shows conditions not found in normal muscles. The muscular fibrillæ are thickened." Phenomena like these seen in myotonia have been produced in animals through veratrin and creatinin poisoning.

The **treatment** is not very satisfactory and the way in which remissions occur is not known.

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MYATONIA CONGENITA

Myatonia congenita was first described by Oppenheim. It is also sometimes spoken of as amyotonia congenita. It is not familial, according to Spiller. Batten has attempted to show that it may be a family disease and refers to two cases in children of one family reported by Sylvestri and three cases in one family recorded by Finkelburg. Cassirer does not believe that it is familial. Certainly it is not generally familial. There is an absence of any familial or etiologic relation with myopathies (Spiller) and a large majority of cases of myatonia are congenital although the condition may not be noticed at birth. In a few cases the condition has appeared rather acutely and reached its height in a few days (Collier and Wilson) and in the case reported by Openshaw symptoms developed after measles.

It is first noticed in these cases that there is a marked atonicity in all or practically all of the muscles of the body. The arms and legs can be over extended and are extremely flaccid. As the child grows older it does not learn to walk, cannot hold up its head and usually cannot sit up. The child generally does not move the limbs, but this is not so much paralysis as atonicity. The tendon reflexes are diminished or absent. In somewhat older children an acute onset of the trouble may suggest the diagnosis of acute poliomyelitis or postdiphtheritic palsy. There is usually no muscular wasting and the condition has a tendency to improve. The generalized atonicity is practically the only symptom present. Sensory changes are absent and the patients are normal mentally so far as can be determined at such an early age. The respiratory muscles may be involved.

Spiller gives a difference between myatonia congenita and muscular dystrophy that the characteristic muscular flacidity is not present in myopathy and the localized muscular wasting is not present in the myatonia. The course of myopathy is progressive and that of myatonia is one of gradual amelioration of all of the symptoms. The myatonia affects the periphery of the limbs especially the intrinsic hand muscles. Myatonia never spreads to parts previously unaffected, but a slow spreading of the affection from muscle to muscle is characteristic of myopathy. The disease is rather closely related to the Werdnig-Hoffman type of progressive muscular atrophy in its clinical characteristics and in fact Rothmann concludes that transition types exist and that a sharp distinction cannot be made between them. According to Spiller, Marburg lists the following differences:

AMYATONIA	WERDNIG-HOFFMAN ATROPHY
Congenital	Acquired early
Usually single case	Familial
General hypotonia	Localized atrophy beginning in pelvic girdle
Atrophy concealed	Atrophy easily recognized
Lost tendon reflex	Tendon reflex in proportion to atrophy
Faradic reaction lost	Reaction of degeneration
Galvanic normal	
Improves	Becomes progressively worse

Spiller summarises the important findings in the muscular and nervous systems in a case examined by him as follows: "The brain was of extraordinary large size for so young a child (25 months), and was as large as that of an adult. Excepting the large size it presented no gross anomaly. The nerve cells in the anterior horns in the cervical and lumbar regions were scarce, and those present were much atrophied. The anterior roots throughout the cord were considerably smaller than in a normal spinal cord and stained imperfectly by the Weigert hematoxlyn method. A peripheral nerve examined was much degenerated. The alteration of the muscle was intense and consisted of greatly atrophied muscle fibers in the midst of other fibers well preserved of overgrowth of sarcolema nuclei and connective tissue nuclei, of excess of fibrous and connective tissues, of increase in size of some muscle fibers and of fatty changes within certain muscle fibers. The intramuscular nerve fibers showed degeneration. The muscle spindles appeared to be normal."

The prognosis in myatonia congenita is usually regarded as good for recovery although in many of these infants their weakened condition makes them an easy prey for infections and but few of them live to become adult.

Gittings and Pemberton found an increase creatine metabolism and Powis and Raper also found a high creatine and relatively low creatinine output. The latter investigators studied the metabolism carefully in a patient four years old. They found fatty stools and an absence of urobilin in the stools leading to the conclusion that there was an acholia. Their patient showed some improvement under treatment with bile salts. Griffiths and Spiller found the liver cells vacuolated but no other pathologic changes have been reported in the liver in this condition.

This is apparently no specific treatment for the affection. A process of upbuilding is of course desirable. The case reported by Chene (quoted by Spiller) in which there was rapid improvement in the myatonia following the recovery from "well-marked rickets" may offer a therapeutic suggestion.

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FAMILIAL DISEASES OF THE SPINAL CORD

Spastic spinal paralysis, uncomplicated, is a very simple symptom complex consisting of a spastic paraparesis with increased tendon reflexes, positive Babinski reflex and nothing else. Theoretically, this syndrome could only be produced by a system degeneration in the pyramidal tracts. Degeneration of the pyramidal tracts is the result of either brain injury or mal-development in the great majority of cases, and in such cases is generally complicated by such condition as mental retardation, aphasias, epilepsy, athetosis, etc. When the pyramidal tract degeneration is secondary to brain lesion the condition is more aptly termed "cerebral diplegia" or is given the name Little's disease. Cases that clinically resemble the syndrome of a primary lateral sclerosis may at necropsy reveal a definite cause as for instance a syphilitic meningomyelitis, multiple sclerosis, etc.

Cases where an exogenous cause for the degeneration of the pyramidal tract is not present are often hereditary. Seeligmüller was perhaps the first to report such a case. Since then a fairly large number of others have been reported by Pelizaeus, Strümpell, Newmark, Erb and others. In many of the hereditary cases a necropsy has shown that the brain was also involved and that the affection was really cerebrospinal although in some cases there were no cerebral symptoms.

The **symptoms** of the trouble may be noticed shortly after birth, but are more often overlooked until the age when the child should walk. In some cases the symptoms have not developed until much later, in Strümpell's case at about 20 years and in Melotti's not until 40 years. According to Jendrassik all the children in the family are usually affected but some may escape. It rarely descends directly from father to son. A study of the family in these cases usually suggests that the condition is due to a blastophoria rather than the expression of an hereditary trait in the Mendelian sense.

The main symptom is the weakness and spasticity usually in the legs, sometimes also in the trunk an upper extremities. The legs can be bent with great difficulty, and when the patient sits on a table the legs at first project forward, then drop slowly often with many interruptions. The extremities may be generally wasted, but there is no localized atrophy. The gait is spastic, often crosslegged from spasticity of the adductors, and in severe cases the patient is unable to walk or even stand. The feet are often contractured to a pes equinus. The tendon reflexes are exaggerated, and there is a positive Babinski reflex. There are no sensory changes, no vasomotor or trophic changes, and the sphincters are not affected.

Theoretically, the **pathological anatomy** should consist of a primary degeneration of the pyramidal tracts alone, but most of the cases that have come to necropsy have shown degeneration also in other fiber systems.

The **diagnosis** is not difficult if a clear family history of the trouble is obtained, but in the case without this the diagnosis must be made from myelitis, multiple sclerosis, spinal-cord tumor, Pott's disease and other conditions affecting the spinal cord. Some of the other familial diseases of the spinal cord such as amyotrophic lateral sclerosis, Friedreich's ataxia, etc., may begin apparently as a pure lateral sclerosis. Syphilitic spastic spinal paralysis must also be differentiated, although it is quite possible that syphilis, especially congenital infections, may be an etiologic factor in the condition.

The **prognosis** of familial spastic spinal paralysis depends largely on the severity of the condition. It is not progressive as a rule, and as the child grows older he usually develops considerably better use of the limbs. The prognosis is worse if the trouble is complicated by cerebral symptoms such as athetosis, epilepsy or imbecility.

The **treatment** directed against the pathologic condition in the spinal cord is practically useless. Even in the case demonstrably of syphilitic origin, anti-syphilitic remedies have little effect. Sedatives and narcotics have no good effect. When the use of the limbs is chiefly hindered by contractures, surgical orthopedics and proper apparatus may be of great benefit (Hoffa). Brissaud and Sicard have suggested injection into the peripheral nerves for the relief of the spasticity, and Foerster recommends the resection of the posterior nerve roots supplying the parts affected. The Foerster operation is physiologically a correct method to relieve the spasticity without causing paralysis. It is a rather difficult and dangerous operation and might be reserved for the more severe cases.

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AMYOTROPHIC LATERAL SCLEROSIS

Probably the first to clearly describe this condition with a definite conception of its pathology was Charcot in 1869 who reported a number of observations of this disease in the next few years. Since then a large number of cases have been recorded in the literature, and Spiller does not regard it as especially rare.

It is not a disease of childhood as a rule. The average age of onset is about 40 years, although Marburg out of 100 cases found as many developing between 50 and 60 years as between 30 and 40 years. Ten cases began between 60 and 70 years. It can begin at any age, cases have been reported from the age of two years on (Kuhn).

Etiology.—For the cases developing in later life a great variety of causes are given. Trauma plays a considerable part in some cases. Hoehl reported a case following an electric shock. Galetta described an emotional origin for one case. The Edinger theory of overstrain apparently explains the development of some cases while Spiller in an earlier paper (1905) considers it an abiotrophy (Gowers, one case). Various intoxications and infections are also described as causes, according to Marburg; lead, arsenic (Williams, Wilson); alcohol, nicotine (Dana); pellegra (Testi); after pregnancy (Dusterwald). A number of cases have been described following acute anterior poliomyelitis. Spiller (1912) suggested that syphilis played an important part in the causation of amyotrophic lateral sclerosis and in two cases with necropsy was able to find round-cell infiltration suggesting syphilis. His findings, however, like those of others seem to show that syphilis is much more frequently associated with disease of the posterior columns and amyotrophy than with amyotrophic lateral sclerosis.

Although not usually considered familial there are a number of instances of a familial tendency on record notably the case reported by Kuhn, Seeligmüller, Testi, Gee, C. H. Brown, Bonardi, Craft, Gordon, Maas, Holmes, Berger, Lent, Luce, Pfaundler, Omerod, Higier, Bruns and Hoffmann. Not all of these cases began in childhood. The sisters reported by Bruns were 23 and 26 years of age. The case reported by Bonardi began at the age of 10 years, Berger's at 12 years, Higier's at eight years. The majority of the familial cases have shown some symptoms when they first attempted to walk.

Symptoms.—The three main clinical features of amyotrophic lateral sclerosis consists of a spastic spinal paralysis usually manifested first and most markedly in the lower extremities, a progressive muscular atrophy which begins usually in the small muscles of the hand and has all of the characteristics of the spinal type of progressive muscular atrophy, and a progressive bulbar palsy. The disease may begin with any one of these features. Usually it is gradual in onset although in a few cases it has been apoplectiform (Schlesinger, Haenel). Both sides of the body are affected but some cases develop at first unilaterally and may remain so for a considerable time. The legs usually present all the phenomena of spasticity, increased tendon reflexes, positive Babinski reflex, etc., but they rarely atrophy. There is often considerable wasting of the legs and there may be intense contractures. There are no sensory symptoms, and the functions of the bladder and rectum are usually not disturbed. The arms are usually held adducted, elbows and wrist flexed, due to the spasticity of the pectoral and flexor muscles. The opponens pollicis and interossei muscles are wasted, and deep spaces appear between the metacarpal bones. The electrical reaction may be simply diminished, or there may be a complete reaction of degeneration. Fibrillary tremors are often present.

Usually, the bulbar symptoms come on only in the end stages of the disease. There is noticed a difficulty in swallowing, a progressive dysarthria, and various paralytic phenomena in the lips, tongue, palate and laryngeal muscles. Rarely the bulbar symptoms usher in the disease. Occasionally there are spastic symptoms in the masseters (masseter clonus, Oppenheim) or even trismus (Schlesinger).

A considerable number of cases have been recorded in which the additional phenomena present cast some doubt upon the diagnosis. The absence of the abdominal reflex in the case reported by Schlesinger suggests a diagnosis of multiple sclerosis (there was an acute onset with bulbar symptoms). The sharp shooting pains described as present in the cases reported by Schultz-Renz, Meyer, and Claude and Lejonne suggest that there was some complicating meningeal irritation. The bladder symptoms described by Sarbo and by Spiller, also the ocular palsies reported by Uthoff, Debore, Strümpell, Sarbo, Spiller and others suggest a diagnosis of cerebrospinal syphilis and a more widespread lesion than is usually present. Spiller's case presented evidence of syphilis on histologic examination.

Mental symptoms are usually absent though increased emotionalism and irritability may be present in the early stages and some dementia later. In most cases, however, the sensorium is remarkably clear.

The familial cases differ in some respects from the ordinary type. There is frequently a congenital weakness even though the child appears well at birth, but they either never learn to walk or else are much delayed. Gee reported a family of three cases. The oldest patient was 37 years of age at the time of his report and had had the trouble all his life. It was very slowly progressive, and even at that age he could use his hands well. His brother and sister were aged 11 and 12 years, respectively, and showed similar signs. Neither had learned to walk until after three years old. While many of these cases are recorded as intelligent, the cases reported by Brown, Higier, Maas and others were demented. Higier's patient also had optic atrophy and Brown's patient an auditory nerve deafness. The other signs are the same as those described in adult cases.

When typical, or well developed amyotrophic lateral sclerosis is not difficult to diagnose, but as the disease usually begins either with the spasticity, the amyatrophy or the bulbar symptoms it is often impossible to decide whether it is not either primary lateral sclerosis, progressive spinal muscular atrophy or progressive bulbar palsy, respectively. If these latter conditions are looked upon as partial forms, so to speak, and due to the same cause or causes of amyotrophic lateral sclerosis, as they probably are, the distinction is not of much clinical importance.

Syringomyelia with the cavity location in the cervical region may cause difficulty for the reason that the cavity may not extend sufficiently into the posterior horn to cause sensory symptoms. Sooner or later, however, the characteristic dissociation of sensation will enable the diagnosis to be made. It should be remembered that syringomyelia is frequently associated with syringobulbia and bulbar symptoms. Pachymeningitis cervicalis hypertrophica may cause all the symptoms of amyotrophic lateral sclerosis, but it is usually accompanied with some pain in the neck, shoulders and arms. Cervical Pott's disease may also be diagnosed by the presence of pain and also by the *x*-ray examination. In children, a cerebral diplegia with pseudobulbar palsy due to bilateral cerebral lesion may simulate amyotrophic lateral sclerosis, but the amyatrophy is not so pronounced or localized, and there would probably be other cerebral symptoms such as athetosis, epilepsy, imbecility, etc.

Pathology.—The pathology present has been carefully studied by Spiller, Rossi-Roussey, Mott, G. Holmes, and others. It may be summarized briefly as follows: The crossed pyramidal tracts and often also the direct pyramidal tracts show marked degeneration which usually extends up to the mid-brain and has been traced by Mott and by Spiller as far as the motor cortex. The spasticity is generally proportionate to the amount of degeneration, but in a case studied by Spiller the greater degeneration was on the side opposite to the greater spasticity. The posterior columns were found degenerated by

Egger, but this was undoubtedly not an uncomplicated case but rather a case of combined sclerosis. Changes have been found in the cells of the motor cortex. The most marked cell degeneration is found in the cells of the anterior horns of the spinal cord. There is also degeneration of the anterior spinal nerve roots. Perivascular infiltration and round cell infiltration of the meninges has been found by Spiller, Czyhlarz and Marburg and others. The muscles show a simple degeneration which is, however, often intense.

No pathologic studies have been made on familial cases.

The **prognosis** in non-familial cases is usually regarded as absolutely bad. In the acute cases death supervenes in about six months or less. The average duration of life is not over two years. I have seen a case of six years duration, and Flourands reports a case of 10 years duration. Crafts reported a case in a child with recovery, but the diagnosis is perhaps a little doubtful. The prognosis in the familial cases is much better for life than in the adult acquired cases, for while they are slowly progressive for a time, they seem to come to a standstill before they arrive at a fatal termination.

Treatment is most unsatisfactory. A cause might be searched for and removed if possible. In cases with a possible syphilitic etiology anti-syphilitic remedies might be tried, but even in cases of spastic spinal paresis known to be due to syphilis, the treatment is not effective. The spasticity may be helped by hot baths, massage and passive movements. In the later stages the bulbar involvement will require tube feeding.

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FRIEDREICH'S ATAXIA

Hereditary ataxia due to a lesion in the spinal cord was first described by Friedreich in 1863. The numerous cases reported since that time have tended to show many variations from the original type, both clinical and pathologic. In 1893 Marie described the cerebellar type. Between these two types there are many examples of transitional forms and also many cases with added signs of other forms or heredodegeneration. Hendrassik classes these cases in one general group and Spiller from the pathologic standpoint points out that most of these cases show pathologic changes not limited to any one part but occurring in various parts of the nervous system, both peripheral and central, as well as in the muscles. As in the case of the muscular dystrophies, there is no sharp line of demarcation between the various types of hereditary ataxia, though the older terms are retained to facilitate description.

Etiology.—An hereditary tendency is a prime factor in the cause of Friedreich's ataxia although there may be no family history of the disease. Friedreich first thought the affection an hereditary tabes until Schultz found a combined degeneration of the posterior and lateral cord. Rutimeyer amplified his description. It is rather uncommon to find direct heredity though frequently all the children in a family will have the disease. Sanger Brown reported its transmission through apparently healthy parents. According to Jendrassik epilepsy and mental disorders are common in the family history of these patients. One of Friedreich's patients had an alcoholic parent.

In a few cases some signs of the affection have been noticed in the first year of life, but the average age of onset is around 12 to 14 years. In the family reported by Brown, the earliest case was six years, the latest 39 years old at the time of onset. Cases have been reported as beginning at the age of 50 years (Jenrassik). There is a tendency for all the cases in a family to begin at about the same age.

Symptoms.—Friedreich in summarizing his cases listed the major symptoms as the ataxia and the speech disturbance and as minor, less constant, symptoms scoliosis, vertigo and nystagmus. He especially emphasized that there was no disturbance of sensation or of special sense. The intelligence was normal, and there were no trophic or sphincter disorders.

Usually the first symptom noticed is the ataxic gait which differs from the tabetic ataxia in being more staggering, falling from side to side, turning about, etc. The position of the body is sometimes peculiar, pelvis forward, shoulders back. The station is also uncertain and is made worse by closing the eyes (Romberg symptom). The legs also show ataxia in the heel to knee test. The ataxia also affects the upper extremities though usually later and to a less degree. There may be only a slight intentional tremor in the hands. Not infrequently there are choreic or athetoid movements in the extremities. Jendrassik also speaks of the occurrence of convulsive tic, especially in the face. Occasionally, the extremities become weak and also there may be an atrophy of the muscles which is most frequent in the small muscles of the hands and feet (Bing, Dejerine, Spiller and others). A peculiar characteristic deformity occurs in the feet (Friedreich's foot). The arch of the foot is very high, the first phalange of the great toe is dorso-flexed and the second phalange is plantar flexed. This is often ascribed to weakness of the muscles or to contractures. Purses has recently brought forward the theory that it is a deformity acquired through the continued effort on the part of the patient to steady himself.

The Babinski reflex is present, but the other skin reflexes are usually normal. Bladder and rectal symptoms are usually absent. In girls menstruation may be delayed in onset.

The tendon reflexes are usually diminished or absent. They have been described as normal or even increased, but these cases were more likely cases of the cerebellar type.

Speech is often affected, being slow and monotonous, sometimes scanning, sometimes explosive and rarely bulbar in type.

Disturbance in mimicry has been reported (Friedreich, Jendrassik) also forced laughing and crying.

Nystagmus is frequently observed, especially on lateral excursion of the eye-balls. The pupillary reaction is generally normal, but the light reaction may be slow and the accommodation reaction preserved (A-R pupil). A slowing of the reaction in accommodation has also been observed. Atrophy of the optic nerves has been reported in a few cases though they are usually normal. The visual field is normal. The special senses are generally normal, and the psyche is not affected. Occasionally there is a spinal scoliosis probably due to some weakness of the back muscles. The extremities are frequently cold and cyanotic.

Sensory symptoms are very rare, all forms of sensation being preserved. This is especially noteworthy in view of the extensive degeneration present in the posterior column of the spinal cord.

Visceral symptoms are also uncommon. Aubertin reported a case with congenital pulmonary stenosis and refers to a similar case reported by Sohrab.

Heart lesions have also been found by Launnois. Spiller reported in one of his cases attacks of vomiting resembling gastric crisis.

Pathology.—The pathology of Friedreich's ataxia has been carefully studied by Spiller, Rutinmeyer, Müller, Gurzetti, Bing, Dejerine and Thomas and others. The most characteristic change is that found in the posterior columns of the spinal cord and, in over half of the cases, also in the lateral columns. It consists of a severe degeneration of the fiber tracts and their replacement with neuroglia which is rather characteristically arranged in whorls or "tourbillons." Other changes in the nervous system that have been recorded are degeneration of the posterior nerve roots and ganglia, degeneration of the anterior horn cells, degeneration in Clark's column, and changes in the dentate nucleus. Spiller and others have recorded marked degeneration in the muscle fibers, when stained by the Marchi method. The muscular degeneration and the muscular atrophy, when it occurs, suggest a combination with a myopathy. Clear cases of the combination of Friedreich's ataxia and a myopathy have been reported by Bing, Camp and others. In many cases there has been observed an appearance of pseudohypertrophy in some of the muscles.

Diagnosis.—Cases conforming in all respects to the clinical picture described by Friedreich would offer no difficulties in diagnosis especially if there was a family history of the affection. Cases complicated by other signs, however, might be extremely difficult to diagnose from multiple sclerosis, juvenile tabes and similar affections. The distinction between Friedreich's ataxia and the hereditary cerebellar ataxia of Marie is discussed under the latter heading. The course of the hereditary ataxia is very slowly progressive and liable to long periods of standstill. Many of these patients live to old age even though the disease began in childhood.

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FAMILIAL SYRINGOMYELIA

Etiology.—The cause of syringomyelia is generally regarded as the breaking down of the gliomatous proliferation near the central canal of the spinal cord. Since this condition is almost certainly due, primarily, to a developmental anomaly it is strange that familial syringomyelia is so rarely reported. According to Haenel heredity plays no rôle in the etiology of syringomyelia ordinarily. The great majority of cases of syringomyelia begin in the cervical region of the cord, but the familial cases have been of the lumbosacral type. The first case of this kind appears to have been published by Bruns in 1903 who found the condition in four out of five children in a family. The father and mother were well. In each case it began at the same age (17 years) with a perforating ulcer of the foot, later trophic changes in the ankle joints developed and gangrene of the toes. There was no dissociation of sensation and no disturbance of bladder or rectal functions. Atrophy below the knee was marked in one of the cases after 10 years duration. The cases reported by Verhoogen and Vandervelde were in two sisters and a brother. In two the hand was the more affected part. One of these patients came to necropsy, and holes were found in Goll's tract and also in the lateral column, but Schlesinger refuses to accept it as syringomyelia. The pathologic findings were certainly not typical. The cases reported by Clark and Grover resembled closely those reported by Bruns. The brother had an affection of the feet from infancy but came under observation at the age of 23 years for painless ulcers on the feet. The sister, age 15 years, began having the trouble at 11 years. An *x*-ray of the feet showed rarefaction of the bones of the foot. There were also sensory changes in the legs of the syringomyelic type, extending half way to the knee. The tendon reflexes were exaggerated, and there was a positive Babinski reflex. There was no disturbance of bladder or rectal function and no scoliosis. Bramanns patients were three brothers, age 13, 11 and 7 years. There was symmetrical gangrene of the fingers and toes and sensory changes. The cases are very briefly reported and suggest also the possibility of Raynaud's disease. According to Schlesinger cases of family syringomyelia have also been reported by Ferranini, Nalbandoff and Preobraschenski, but their original reports are inaccessible to me.

Syringomyela, in such cases, must be differentiated from instances of spina bifida and malformations of the sacrolumbar region of the spinal cord which are not uncommon. Spina bifida occulta will, of course, be diagnosed by *x*-ray examination.

The prognosis is bad for recovery, but the condition seems to be very little progressive. Amputation of the foot to be rid of the gangrenous part is usually followed by prompt healing and a useful stump.

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FAMILIAL DISEASES SIMULATING MULTIPLE SCLEROSIS

A certain number of cases have been reported as cases of familial multiple sclerosis or as a familial affection simulating multiple sclerosis among these we may include those reported by Moncorvo, Stieglitz, Moussons, Dreschfeld, Pollard, Railton, Cestan and Guillain, Bernhardt, Pelizaeus, Higier, Eichhorst, Westphal, and Friedman.

In a disease with such protean manifestations as multiple sclerosis the diagnosis is often extremely difficult, and the actual occurrence of familial multiple sclerosis is denied by many authors.

In analyzing the above cases one finds that those reported by Moncorvo and by Friedman were apparently of syphilitic origin. Both were so diagnosed, and both improved under anti-syphilitic treatment. Syphilis is apparently not an etiologic factor in multiple sclerosis but, as Spiller and Camp pointed out, the diagnosis between multiple sclerosis and disseminated cerebrospinal syphilis may be impossible from the clinical symptoms alone, and only by the biologic tests can the diagnosis be definitely established. There is no question but that many cases of multiple sclerosis in adults are mistaken for cerebrospinal syphilis and *vice versa*, and the same applies to children with congenital syphilis.

In the case reported by Pesker the symptoms were convulsions in the first year, difficulty in speech and gait at six years, increasing paraplegia, contractures, intention tremor of the hands, horizontal nystagmus, slow and scanning speech and diminished intelligence. A brother died who had exactly the same symptoms, and at necropsy there was found defective development of the spinal cord, especially in the direct cerebellar and Goll's column, but also in the crossed pyramidal tract. Clark's column was degenerated. There was no change in the blood-vessels, but the cerebral cortex was also affected, and there was a marked accumulation of small round cells. In discussing this case Marie refers to the case of Pelizaeus as exactly similar and suggested that it be regarded as a new form of a family affection. A syphilitic etiology was not suspected, but the round-cell infiltration of the cortex would suggest it to neuropathologists of the present day, Pelizaeus regarded his case as one of multiple sclerosis, but the diagnosis is open to question. The cases reported by Dreschfeld and by Railton were clearly cases of cerebral diplegia with athetosis, and the case reported by Bernhardt was a spastic paraplegia of unknown origin. The cases reported by Stieglits and by Pollard were not familial or hereditary. Eichhorst related the case of a woman, age 36, who had three children. Two years before the birth of the third child she began to complain of weakness in the legs which eventually developed into the symptoms of multiple sclerosis The diagnosis was confirmed by necropsy. The third child at the age of eight also developed multiple sclerosis and died three months later. The diagnosis was also confirmed by necropsy. This is apparently the only case in the literature of hereditary multiple sclerosis with the diagnosis confirmed by necropsy. It apparently did not affect the other children in the family. Higier reported four sisters who began having symptoms of stiffness and weakness in the legs at ages varying from 7 to 12 years. There was a negative family history, and they were normal at birth, and up to the time of the development of these symptoms. All had a positive Babinski reflex and developed contractures in the legs so that walking was impossible. Two of the sisters developed intention tremor in the upper extremities; two of them monotonous, scanning speech. Primary optic atrophy was present in all four sisters with disturbance of the visual field and, in one case, bitemporal hemi-anopsia. The oldest and youngest sisters showed mental defect. The author considers the diagnosis of familial multiple sclerosis but rejects it largely because of the absence of any remissions. He prefers to regard them as examples of family cerebral diplegia such as was first described by Sachs, but calls attention to certain differences from ordinary cerebral diplegia such as the insidious onset, chronic progressive course, familial tendency, the beginning in childhood, the absence of any apparently etiology and the absence of any evidence of cortical irritation.

It seems probable that practically all the cases in the literature of so-called familial multiple sclerosis are cases like those of Higier, Pesker's being the only one with necropsy.

The prognosis for any recovery in these cases is bad for their course is slowly progressive, at least, to a certain stage when it may become stationary. Practically all that can be done is to minister to their comfort and provide them with something to occupy their mind.

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FAMILY PERIODIC PARALYSIS (FAMILY MYOPLEGIA)

This rare condition was first described by Hartwig in 1874 and in 1902 Oddo and Audibert were able to find 64 cases recorded in the literature. A considerable number have been recorded since that time. Holtzappel in 1909 recorded 17 cases in one family under his observation. According to Oppenheim, cases have been reported by Romberg and by Cavere that resemble periodic paralysis but which were due to malaria and cured by the administration of quinin. The family cases have no apparent relation to these cases except for some clinical resemblance.

Most of the diseases of the spinal cord classed as familial have for a distinguishing characteristic a slowly progressive course. The disease under consideration comes on in attacks which usually began in childhood or youth (age 10 to 20 years) and recur at intervals through life.

The attacks generally begin at night, the patient awakening to find himself completely paralyzed. The cranial nerves usually escape although Singer reported ptosis and Mitchell and also Fuchs report dysphagia at the height of the attack. If awake the patient may have a short warning of the attack by way of some tingling through the body and often a sensation of thirst or occasionally an outburst of sweating. The attacks very rarely come on in daytime, but usually after sleeping or resting. Attacks have been reported as caused by excessive exertion or by overeating. Partial attacks may occur in which only part of the body is affected, usually the legs and feet, or the premonitory symptoms may appear without the attacks following. Holtzapple's patients called this "feeling the attacks."

The paralysis is flaccid and does not come on instantaneously but in the course of a few minutes. The tendon reflexes are lost as a rule (in Bornstein's case the tendon reflexes were increased, but the skin reflexes lost) and the myotatic irritability is also lost. In the muscle there is no reaction of degeneration, but the muscle does not react at all ("Cadaveric reaction"). The electric reactions return to normal with the return of power, which is generally gradual. The average duration of the attack is about 12 hours, but the time varies anywhere from one-half hour to a week or more. The interval between attacks is also very variable in length and during this interval the patient feels well. During the attack the patient is perfectly conscious. There are no sensory changes and usually no disturbance of the sphincters though there may be some difficulty in emptying the bladder on account of the paralysis of the abdominal muscles. There is usually difficulty in coughing or clearing the throat.

Oppenheim and several others have observed dilatation of the heart accompanied by signs of mitral regurgitation during the attacks, but in the interval all heart signs are absent.

Goldflam and others have examined pieces of muscle obtained at biopsies and found hypertrophy of the muscle fibrillæ, vacuoles in the muscle fibers and a glassy appearance. They would favor regarding it as a muscle disorder. Bernhardt reported a case combined with progressive muscular dystrophy, and the case reported by Oppenheim also later developed a dystrophy. Oddo also takes the view that the condition is a congenital malformation of the muscle tissue but considers it more in relation to myotonia congenita.

Attention has also been called to its relation to epilepsy (Higier) but the transient paralysis occurring either as an equivalent or as a sequel of an epileptic attack is quite different in its nature. Migraine was found by Holtzapple to be very frequent in the family he reported. Out of 30 affected members 17 had periodic paralysis and 18 had migraine, five of them having both. The theory has been advanced that periodic paralysis may be due to a vasomotor disturbance in the anterior horns of the spinal cord. This, of course, is entirely theoretical.

The nature of the attacks also suggest a toxic origin, but metabolic studies have not given much result. Goldflam and also Schlesinger found albumen in the urine after attacks and Goldflam, Singer and several others speak of an increase in the toxicity of the urine.

The diagnosis is not difficult if the family history is obtained. In no other affection does one find the same type of electrical reaction. An hysterical condition might be thought of, but hysterical palsies are more in the nature of contractures, inhibitions, etc. There are usually sensory changes, hysterogenous zones, etc., and no loss of reflexes or electrical reaction.

The prognosis is generally good for the attack. The only deaths reported in attacks were the six reported by Holtzapple one of which occurred in his presence. He states that the patient was occasionally having some difficulty in breathing but was mentally clear when he suddenly gasped, his pulse became irregular, and he died apparently because of an acute cardiac dilatation. Since these patients are unable to cough or raise sputum an attack occurring coincidently with a bronchial or lung affection would probably be serious. The attacks will likely recur through life and muscular dystrophies may develop.

Attacks are said to have been aborted by giving large doses of bromid and caffein, but too strong drugs should not be given. In the interval attention should be paid to the diet and general hygiene with especial avoidance of constipation and fatigue.

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HEREDITARY CEREBELLAR DISEASE

Hereditary Cerebellar Ataxia.—This condition was first described by Pierre Marie in 1893. He was able to collect 16 cases in two of which there was an opportunity to examine the nervous system. The marked atrophy of the cerebellum in these cases and the fact that the knee-jerks were increased instead of diminished or lost as in Friedreich's ataxia were the chief points of distinction from that condition. Later investigations have shown that there are all degrees of transition between the two conditions. In the same family both types of ataxia may be observed. As a rule the cerebellar type develops a little later in life than the spinal although the cerebellar type may begin in childhood.

As in the spinal cases the ataxia is usually the first symptom noticed. It may be purely cerebellar in type with dysmetria, asynergia and other cerebellar localizing signs. The ataxia affects the upper as well as the lower extremities, and may be accompanied by an intention tremor (Bruns). Nystagmus and the disturbance in speech are the same as those found in Friedreich's ataxia. The nystagmus is less regular and has a slower rate than that of multiple sclerosis. Extra-ocular palsies and ptosis may occur. An argyl-Robertson pupil has been observed and Dercum has reported a case with "reversed A-R pupil," i.e., the pupil reacted to light, but not in accommodation. Optic nerve atrophy is not uncommon. Both optic atrophy and extra-ocular palsies are very rare in Friedreich's ataxia. The tendon reflexes are increased and there may be an ankle clonus though in a late stage of the disease both many be absent. Bladder symptoms are more frequent than in the spinal type. Mental symptoms are not so infrequent in cerebellar cases. The deformity of the foot so characteristic of Friedreich's ataxia and attributed by Bruns to an atrophic paralysis is rarely seen in cerebellar ataxia.

Pathologic investigations show that only rarely is the anatomic change confined to the cerebellum. Bing found only one case out of nine diagnosed as hereditary cerebellar ataxia that had no lesion except in the cerebellum. Twice were there lesions both in cerebellum and spinal cord, once in cerebrum and cerebellum and in the rest chiefly spinal. The cerebellar lesions are for the most part simply aplasia.

The course and prognosis of hereditary cerebellar ataxia is the same as that for Friedreich's ataxia. The progress of the affection is slow and it frequently is stationary for years. It does not shorten life, but there is no cure.

The differential diagnosis between hereditary cerebellar ataxia and Freidreich's ataxia has been considered in connection with the description of the symptoms of the former. It is frequently impossible to arrive at a definite conclusion. The diagnosis of hereditary ataxia from other disorders of the nervous system is more difficult when we are dealing with the cerebellar type than when the spinal type is present. The cerebellar type may closely resemble multiple sclerosis which, while rare in children, has been described and has also been described as familial in exceptional cases. The course of multiple sclerosis is usually different; it may be sudden in onset, have periods of remission or acute exacerbations, etc., while the course of hereditary ataxia is very slowly progressive. In multiple sclerosis a paraplegia or hemiplegia may occur and disappear, in hereditary ataxia the symptoms remain. Optic atrophy may be present in either, but optic neuritis only occurs in multiple sclerosis. Multiple sclerosis frequently gives rise to bladder disturbance, sensory changes, disturbance of special sense organs, amaurosis, multiple scotomata, etc. Familial diseases simulating multiple sclerosis such as cases reported by Bernhardt, Dreschfeld, Pelizaeus may give even more difficulty. Their diagnosis is discussed under that heading. Bruns records a case in a child with "spastic paresis, intention tremor, ataxia, nystagmus, speech disturbance and optic atrophy." The necropsy showed "softening in the region of both precentral convolutions following thrombosis of pial veins." Oppenheim points out that an acute encephalitis located in the cerebellum might give the signs of cerebellar ataxia especially if the etiology of the affection was congenital lues. A tumor of the cerebellum might also give the same symptoms. The diagnosis could only be made by the discovery of other evidence of tumor—headache, choked disc, etc.

No treatment so far recommended has been beneficial in hereditary cerebellar ataxia. Re-educational exercises for ataxia (Frenkel) cannot be expected to benefit for the reason that the condition is not due to sensory loss.

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CHAPTER CLXXII

CONVULSIONS IN INFANCY AND CHILDHOOD

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Definition.—A convulsion is defined by Hughlings Jackson* as "a sudden discharge of many nervous arrangements representing movements at once or nearly together, because the cells subserving such movements have become highly unstable." A convulsion then is an expression of an acute, subacute, or chronic change in the body, due to some abnormal condition. It is not a disease but a symptom of disease or altered state of the body.

History.—A history of convulsions would really be a history of medicine; from the time of Hippocrates medical men have turned their attention to this subject. At the beginning of the eighteenth century, Baumes, professor in Montpellier, published a monograph on convulsions, which he described under the names of eclampsia and epilepsy. He saw only a difference in duration and course between the two, the former being an acute affair, the latter a chronic one.

Brachet† published a handbook on convulsions in childhood in 1824 and a second edition in 1835. In 1834, Zaugerl‡ published a monograph in Vienna.* Both these authors separated eclampsia from epilepsy. After this many articles appeared, such as Duge's "Eclampsia in Children in the First Year in Relation to Apoplexy and Tetanus."§

In England, Doctors Good and Locock were early writers on convulsions. In Germany, Henke and Meissner separated eclampsia and epilepsy. E. Barthez and F. Rilliet were the first to differentiate convulsions due to a lesion of the central nervous system and those apparently free from any pathologic change in the organism.

Clonic convulsions were first described in France by Dance, who observed them in adults. Tonnette‖ described them under the name of "A New Convulsive Disease in Infants," and a month later, Constant published notes of the same disease with contractures in any and all muscles, really an early description of tetany.

Kinds of Convulsions.—From a clinical standpoint the types of convulsions are divided into clonic and tonic. In clonic convulsions there is contraction and relaxation of a group or groups of muscles of the body, more or less rapid, which later may, either by exhaustion or great rapidity, resemble tetanus (tetanic contraction). Tonic contractions are really not convulsive movements at all: There is rigidity of a group or groups of muscles continuing for minutes, hours, or days, really a constant spasm as in tetany. This may occur in one group of muscles, as in glottic spasm. There may be a combination of both tonic and clonic convulsions. In older children, usually after the third year, there is a form of convulsion with disturbed consciousness, a few incoördinate movements, and some disturbance of speech, but not muscular contraction—the so-called psychic equivalent of convulsions (petit mal). Another type of convulsive movements may occur in the form of attacks without loss of consciousness, simply an exaggeration of normal movements, as in spasms, rotatorius, spasms nutans, and eclampsia nutans.

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* From Leonard Guthrie, in "Diseases of children," edited by Gunod. Batten and Thursfield (1913).

† Mémoire sur les causes des convulsions chez les enfants et sure les moyens d'y remédier.

‡ Über die Convulsionen im Kindlichen alter Wien.

§ Mémoire de l' Académie de Médicine.

‖ Gaz. med. No. 1 (January, 1832).

The so-called "inward convulsion" of the laity is either a true general tonic convulsion or in reality only a chill as seen so frequently in young children and babies; they become pale, slightly rigid, may or may not lose consciousness; in children, the teeth do not usually chatter.

Physiologic Anatomy of Coördinate and Incoördinate Movements.—In order to understand the entire significance of a convulsion one must ask oneself, first, how does the attack begin, where does it start from, and what takes place there; second, what has been going on in the body and the central nervous system previous to the attack. While it is impossible to answer these questions completely at the present time, there are many facts of interest.

In answer to the first question, it has been shown that the medulla and pons as well as the cortex of the brain can be the starting point of convulsions. Kussmaul proved that by cutting through the brain above the pons varolii, general convulsions may ensue. It is well known that in anencephalic monsters general convulsions may occur. The pons as an area of motility has been demonstrated by Nothnagel and later by Luce. The former has described a convulsion center there. Piercing the substance of the pons varolii in the rabbit, causes typical epileptiform convulsion (Bechterew). Nothnagel tried to localize this center in the floor of the fourth ventricle as extending upward over the ala cinerea to the corpora quadrigemina region; mesially it comprised the eminentia teres, but laterally the boundary was more difficult to define, but surely did not extend over the locus ceruleus and the tuberculum acousticium. These convulsions, however, are probably due to some secondary stimulation of the cortical centers, for when the hemispheres are removed, only tonic and not clonic convulsions occur. Thus there is most likely no real center in the pons, but injury causes some secondary irritation of the cortical centers. It is well known that electrical stimulation of certain portions of the cortex will cause definite motor phenomena. Most of these so-called motor points lie in a circumscribed area in the gyrus centralis anterior, to a certain extent in neighboring gyri, and in the operculum. The stimulus is taken up by cortical and association fibers and carried to the centrifugal neuron. This is shown by treating the cortex with cocain, when the reaction will not occur (Monakow).

If the electrode is thrust deeper into the brain, or the cortex is removed and the deeper portions stimulated, a reaction will occur it is true, but only for a short time, after which it is impossible to evoke a response. The stimulus goes from the cortex through the internal capsule to the anterior horn cells of the spinal cord and thence to the muscles. The course from the internal capsule to the anterior horn cells is but partly known. The pyramidal tracts are the main paths but the intraspinal bundle of Monakow, and other so-called extrapyramidal tracts, also help in the conduction.

If the cortex is stimulated at one point, let us say the right face region, the muscles of the face on the left side of the body begin to contract. If this stimulation is increased or continued the arm muscles and finally the leg muscles on the left side also become involved. Still further stimulation causes an involvement of the right side of the body. If a curve is made of the muscular contraction, caused by an electrical stimulus to the cortex, we get a picture showing that the contraction continues long after the stimulus has stopped. There is first a tonic contraction, then a continuation of the contraction, but of greater intensity, and finally long after the stimulus has been removed, the clonic contraction begins.

Luciani believes that only in the cortex general convulsions can originate, and a typical epileptiform convulsion can only occur with intact motor area. Ziehen and Binswanger, however, think that the subcortical centers in the pons and medulla may well be the starting point of general convulsions. Monakow believes this possible from the experiments of Prus, who showed that stimulation of the posterior corpora quadrigemina in dogs produced clonic convulsions, and that by stimulation of the anterior corpora quadrigemina he could produce tonic convulsions.

According to Monakow the procedure in the production of convulsions is somewhat as follows: The starting point of the attack is in the cortex and in the motor area, usually in an extremity zone. The stimulation goes through the internal capsule (projection fibers), through the pyramidal tract, and the other corticofugal fibers to the hind brain (substantia nigra), to the tegmentum, then to the pons floor and the medulla oblongata (the extra pyramidal fibers), and then to the anterior horn cells of the cord. Before normal coördinated muscular as well as convulsive movements can occur, there must be a participation of stimuli (or impulses) from (*a*) scattered solitary and groups of cells forming the formatio reticularis (in the medulla); (*b*) the cells in the substantia nigra; (*c*) the cells from the pyramidal tracts, and finally, various portions of the cerebellum. Typical epileptiform convulsions, beginning with tonic and ending in clonic spasm, can originate only in the cortex. The highest form of epileptic attack is the cortical one; the lowest form is the so-called spinal epilepsy represented by intermittent tonic contractions as produced at times by severing the cord at a high level.

The second question "What is going on in the body before the convulsion occurs," is really a broad consideration of the etiologic factors. If no lesion in the spinal cord and brain demonstrable with the methods at present at our command is produced, we term the convulsions functional. However if there is a pathologic condition, such as inflammation of the brain, or meninges, spinal cord tumors, hydrocephalus, and so forth, we call the convulsions organic. This classification is useful only from a clinical standpoint; as our methods of examination of the nervous system become more accurate, as our knowledge of the physiology and chemistry of the body grows greater, we will be able to restrict the exact meaning of these terms more and more. We must not forget when we say the convulsions are functional, that this does not necessarily signify that the body is in a normal condition previous to the outbreak, for as is illustrated in tetany, there is a distinct pathologic variation in the irritability of the nervous system to galvanic stimuli.

In general, and for clinical purposes, four general conditions may be said to bring on convulsions at any age. First, the spasmophilic diathesis; second, epilepsy; third, a group without special change in the central nervous system, due to poisons directly introduced into the body, such as lead, strychnin, morphine; the toxins of the infectious diseases, and abnormal chemical processes in the body where the cells themselves are stimulated; fourth, pathologic lesions in the brain and spinal cord, which, by direct or indirect effect on the brain and cord, cause the convulsions. Whether cyanosis and increased CO_2 in the blood can be a cause of convulsions may be doubted. Finkelstein has pointed out the rarity of convulsions in laryngeal diphtheria. Whether reflex causes (eclampsia reflectoria) such as foreign bodies in the ear and the nose, teething, phimosis, worms, and so forth, can cause convulsions, is still an open question. These etiologic factors will be dilated upon at greater length as we get to the various diseases which may be the immediate cause of the convulsion.

The age of the child is so important a factor that we will discuss the different causes of convulsions in various age groups.

CONVULSIONS IN THE NEWBORN

Convulsions in the newborn may be, first, functional or toxic in origin, second, bacterial (really toxic), and third, organic. During the newborn period convulsions due to organic causes are much more common than those due to functional conditions. In fact the functional causes must be looked at with great suspicion during this period of life. However, they must be borne in mind so that absolutely bad prognoses are not made in every case of convulsions in the newborn. This increased proportion of organic causes of convulsion in the newborn as contrasted with the reversed condition in the older child is due to the greater opportunities for grave injury and hemorrhage within the head and to the prevalence of congenital defects manifesting themselves at this age. Why functional conditions should be less prevalent at this time of life than in older children (tetany being practically unknown before the second month and then rare between the second and sixth months) was supposed by Soltman to be due to the diminished irritability of the cortex and the peripheral nervous system on account of the undeveloped condition of the medullated nervous fibers and a lessened irritability of the cortex to stimuli.

The character of the convulsions are most frequently clonic but may be tonic, especially if conditions occur in the posterior fossa, causing irritation of the lower nuclei. At times both forms are present. The frequency of convulsions in the newborn is shown by the following table:

INCIDENCE OF CONVULSIONS IN THE NEWBORN

Dr. Porak et Duranti, Paris
Maternité Clinique d'accouchment,
(1900–1901), 5078 cases.

	Cases	Convulsions	Death	Incidence
Full term	5078	10	6	0.19
Premature	630	17	14	2.69
Dr. Maygier, Paris Charité (1898–1901)	2885	4	1	0.13
Dr. Charpentier de Rebes, Paris Hôtel, Dien (1898–1901)	4321	9	1	0.18

Functional Causes of Convulsions in the Newborn.—(1) Eclampsia Neonatorum; (2) gastro-intestinal; (3) reflex, especially in the premature; (4) mineral poisons such as lead, morphine, atropin, alcohol, etc.; (5) respiratory conditions inducing cyanosis, such as marked nasal obstruction, atelectasis, congenital cardiac disease, etc.

1. Eclampsia Neonatorum.—The word "eclampsia" is loosely used here and must not be confused with other general convulsions. The condition is one occurring in newborn infants whose mothers have been suffering from eclampsia either during pregnancy, or labor, or post-partum. These convulsions are, according to Halbin, an expression of the toxic substances from the mother's blood transmitted by means of the placenta into the child's circulation and reproducing the mother's condition. The nature of these toxic substances in the maternal blood is as yet unknown. Many children of such mothers are born dead or in poor condition and die soon after birth; others have a hemorrhagic tendency, and yet others have general convulsions.

Symptoms.—The convulsions start during the first few days of life, usually on the first day. They are clonic in nature. The face is almost always first involved, though the extremities may also be the first. The convulsions may last moments or hours and may recur very frequently. The children become cyanotic due to contraction of the thoracic musculature. The convulsions rarely last more than two or three days and the child may recover, or the convulsions increase in severity and frequency and the child dies. The severity of the disease in the mother is no indication of the condition in the child. The pathologic anatomy according to Schmorl, Lubarsch and Deinst reveals thrombosis in the various viscera, degeneration of the parenchyma of the viscera, fatty degeneration or necrosis in liver and kidney, subpleural, subepicardial, and endocardial hemorrhage. According to Stumpf convulsions may occur in utero. The prognosis for the life of the child is usually bad, but not necessarily so. Schneider found that in 34 cases of maternal eclampsia there were:

37 children born;
15 spontaneously—12 alive—3 dead;
12 with forceps—11 alive—1 dead;
10 extracted—3 alive—7 dead;
37 cases—13 dead—about 35 per cent. mortality.
Only one of the cases had convulsions.

Lichtenstein—of 371 children from eclamptic mothers
250 born alive;
121 born dead;
23 died later.
Mortality 38.81 per cent.

Diagnosis.—The history is usually sufficient. However acute syphilitic ependymitis with effusion into the ventricles may stimulate the condition, and inasmuch as lumbar puncture is of great value in this condition, lumbar puncture should always be done in cases of eclampsia neonatorum, providing the convulsions are very frequent and of any duration.

Treatment.—According to Halbin's theory, toxic substances floating in the blood are the exciting cause in the symptoms in the newborn. It is natural to suppose that these substances may be excreted in the mother's breast milk and thus increase the amount of toxin above that already brought to the child's blood. It seemed proper, therefore, to avoid giving the breast milk for a few days, having the breast artificially emptied and the milk thrown away. After three to five days the baby is put to the breast.

Experiments with this milk upon guinea pigs and rabbits by the author have shown that the milk does not act differently than milk from normal women. Von Reuss also showed that this milk does not produce toxic symptoms. However, it is simple enough to give the baby the benefit of the doubt and feed a simple milk mixture and later give the mother's milk. Lumbar puncture for the purpose of abating the severity of the convulsions and for diagnostic purpose should be done if the convulsions persist longer than 24 hours.

2. Gastro-intestinal.—That convulsions in the newborn due to the gastro-intestinal tract similar to those of older infants and children may occur, we are not prepared to disprove, but we should venture to say that they are very rare. This part of our subject is difficult and hard to dilate upon for accurate data is not available. We would caution the medical man to give a very carefully considered prognosis in these cases. I refer to the newborn infant after the third or fourth day of life, often in the second week. The child begins to have some temperature and then repeated general clonic convulsions, the entire picture lasting three to seven days but the convulsions rarely extending over two to three days. I have treated these cases with or without cathartics and irrigations of the colon, by withdrawing the breast or bottle, or by not doing so, without much apparent effect. The spinal fluid is normal in its cytology and chemistry, but usually greatly increased in quantity and pressure. It is not often that we stop the nursing and at best only for 24 hours. One favorable prognostic observation is that the ability of the babies to nurse does not seem to be abated. They regularly get well. Cow's milk idiosyncrasy may be considered, but most of our cases occurred in the breast fed and not during the first few days of life. If the child is on a cow's milk mixture it might be advisable to stop this, and give breast milk. The less catharsis and bowel irrigation done to newborn infants the better. Starvation (barley water or plain water diet) should not be extended over 24 hours.

3. Reflex Convulsions.—These have been reported to occur in the newborn, especially in the premature. Each reports a case of a child which had convulsions whenever removed from the warm room into a cooler atmosphere.

4. Convulsions due to poisons such as lead, alcohol, atropin, strychnin, etc. will be discussed later under convulsions in older children. Soltman reports a case of convulsions in the newborn whose mother was an alcoholic; alcohol was demonstrated in the milk.

5. Cyanosis.—In cases of asphyxia pallida, congenital heart disease, or atelectasis, we have seen convulsions occur. These may have been due to the cyanosis, or the cyanosis may have been a result of some other condition or the result of the convulsion. Finkelstein doubts very seriously that cyanosis can produce convulsions, pointing to the fact that cases of laryngeal diphtheria with marked cyanosis rarely have convulsions.

Bacterial Causes.—Bacteremia with various types of organism may cause convulsions. As is well known many of the usual and unusual organisms may produce a bacteremia in early life, such as the hemorrhagic streptococcus, ordinary streptococci, coli, staphylococcus, pyocyaneus, pneumococcus, etc. These cause bacteremia with or without a meningitis or an encephalitis. Other signs of sepsis are present, including temperature, petechiæ, etc. Lumbar puncture should be done and treatment of the underlying conditions begun.

Tetanus may be considered alone on account of the specific type of convulsion and because here there is really no bacteremia but a local infection and a secondary toxemia. The etiologic factor, the bacillus of tetanus, may gain its entry into the body at any point, but the umbilicus is a favorite one. For a description of the bacillus of tetanus the reader is referred to any good textbook of bacterology. The incubation period depends somewhat upon the portal of entry of the bacillus and its proximity to the large nerves. The toxin regularly travels by means of the nerve stems and symptoms are not produced until the toxin reaches the spinal cord. In a report on "Tetanus in Soldiers" to the British War Office, Maj. Gen. Sir David Bruce* gave the average incubation period as 39 days. This in the larger body of an adult is understandable in the light of Meyer Halsey and Ransom's† work. They showed that the larger the body the longer the incubation period.

The symptoms of tetanus in the newborn may better be found in the section on diseases of the newborn, but in brief it may be stated here that symptoms do not usually begin until the end of the first week or during the second week of life: If the convulsions are tonic in character, the muscles of the face are first affected so that nursing becomes difficult. Attempts at forcing the nipple into the child's mouth cause contraction of the masseters, the forehead is wrinkled, the mouth drawn and the face takes on the characteristic look (Risus Sardonicus). Soon the convulsions become general with periods of remission. Any noise, or any disturbance of the infant brings on the attack. The duration is from one or two days to a few weeks. The prognosis is bad, about 50 per cent. or more dying according to different authors—the longer the duration the better the prognosis. The treatment consists of early and repeated injections of tetanus antitoxin intraspinally, intravenously, and subcutaneously.

The organic causes of convulsions in the newborn are many and may be divided into two great groups. First, those due to trauma and hemorrhage; and second, those due to congenital causes. These in general are porencephalus, hydrocephalus, microcephalus, brain hypertrophy, syphilis of the nervous system, acute encephalitis or leptomeningitis, thrombosis, marantic or phlebitic, and affections of the bulbar nuclei.

Porencephalus may be congenital or the end result of cerebral hemorrhage or of an encephalitis. A diagnosis cannot usually be made; but constantly localized right or left sided convulsions occurring soon after birth, may lead one to suspect it. In addition, Bokay's light test, transluminating the skull anteriorly and posteriorly, and from right to left, might give a clue. Lumbar puncture is of no help.

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*London Letter, J. A. M. A., lxiii, No. 7, p. 546.

†Meyer, Halsey and Ranson, Tetanusstudien, Jaffe's Festschrift, Brunswick, (1901).

Hydrocephalus does not usually give rise to convulsions in the first few days of life. The constant increase in the size of the head, the circumference of the head as compared with the circumference of the chest, the separation of the sutures, the position of the eyes and the nystagmus, may be guiding features. Lumbar puncture here is of some use and should be done at regular intervals according to the rapidity of the increase in the measurements of the head.

In microcephalus the circumference of the head is usually smaller than that of the chest, the convulsions are frequently tonic instead of clonic, there is considerable rigidity, the sutures are closed, and the fontanel is small. Lumbar puncture is palliative.

Cases of true brain hypertrophy with enlargement of the head have been reported with convulsions. The diagnosis is impossible during life.

Syphilis of the nervous system in the newborn does not usually give symptoms. Occasionally hydrocephalus is on a syphilitic basis, especially if there is an acute effusion into the ventricles. Here lumbar puncture is of great use. Meningitis syphilitica, myelitis, and encephalitis may occur; a cell count of the cerebrospinal fluid will be of service in making a diagnosis. A lymphacytic cell increase may be present. The Wasserman reaction, as is well known, may be absent in both the blood and the spinal cord fluid at this time of life.

Pachymeningitis hemorrhagica interna is usually without symptoms in the newborn, but may give an effusion between the dura and pia with general convulsions (Doehle).

Acute encephalitis and leptomeningitis secondary to sepsis, with high fever and general convulsions, sopor and opisthotonus, is a familiar result of sepsis in the course of a general bacteremia in the newborn. Acute encephalitic disease in the newborn period is becoming more frequently recognized—Virchow's contention that the signs of fat infiltration in the cortex is a result of an encephalitis is being substantiated.

Marantic and inflammatory thrombosis are often accompanied with convulsions. An examination of the eye-grounds helps at times. In the former (marantic) there is usually stupor and a marasmic condition. In the latter there are often tonic convulsions; the cerebrospinal fluid varies from a brown to a green color.

Convulsions due to involvement or absence of the bulbar nuclei are due either (1) to direct involvement of the nuclei, or (2) to indirect involvement, through pressure. In the first group there is icterus of the nuclei without general icterus (Schmorl and Beneke). The children are seen on the first day of life with tonic convulsions and bulbar symptoms. There is a familial tendency. In the second group, due to indirect pressure, Esch describes a case with subtentorial hemorrhage compressing the nuclei, with the convulsions mostly tonic; Seitz describes another case with infratentorial hemorrhage, which after 42 hours had cyanosis and apnea, spasm of the extremities, opisthotonus and then clonic convulsions.

Convulsions in the Newborn Due to Fracture of the Skull.—Fractures of the vault may be single or multiple. They may not cause any trouble at all, or they may produce an external or internal cephalhematoma. One should be very careful in making a diagnosis of fracture, for the ridge raised by the periosteum in the case of an ordinary external cephalhematoma is easily mistaken for a fracture. The fissures are often not discovered until autopsy. A form of fracture that is of importance is the one that tears the dura and meninges, producing a hernia cerebri—so-called meningocele spuri traumatica. This may grow larger in size and cause general convulsions. Fractures at the base of the skull are very rare. They cause bleeding into the medulla with general convulsions and respiratory symptoms.

Convulsions Due to Trauma and Hemorrhage. Contusio Cerebri.—In severe labors with marked asphyxia, there are at times both tonic and clonic convulsions which would indicate a hemorrhage, however, none is present and the brain shows edema and hyperemia. Cerebral hemorrhage, or better, intra-cranial hemorrhage, is a relatively common cause for convulsions. However, if one is fortunate enough to secure autopsies in these cases, one realizes quickly that every case of convulsions is not due to cerebral hemorrhage, that the diagnosis is more frequently made than is warranted, and that one is very often surprised at the absence of a cerebral hemorrhage. Thus in a case seen recently, the child was born after a very severe instrumental labor, with a facial paralysis which proved to be peripheral and due to pressure of the forceps. On the third day the child had severe general clonic convulsions and refused to take its food. Convulsions occurred every three or four days, the child dying on the seventeenth day. Post-mortem showed a perfectly normal brain, but a large adrenal hemorrhage.

Hemorrhage may be epidural (cephalhematoma interna), subdural, between the dura and the arachnoid, or sub-arachnoid, on the surface of the brain. The epidural hemorrhages usually small, cause very little or no trouble, and are not easily diagnosticated. The subarachnoid hemorrhages cover the surface of one or both hemispheres. They are rarely one-sided, usually involving one or the other motor area, causing general convulsions with hypertonia and disturbances of the sensorium. The subdural hemorrhages are due to compression by means of forceps, tearing of the falx cerebri, or the tearing of veins leading into the sagittal sinus. They may, however, occur in easy births, as in the premature.

In general, cerebral hemorrhages may be divided into supratentorial, infratentorial, and mixed.

Supratentorial hemorrhage causes restlessness, bulging fontanel, and increase in the width of the lambda suture on the side of the hemorrhage; pallor, unilateral facial paralysis, paralysis of the hypoglossis (deviation of the tongue) and of the accessorius (head held to the side of hemorrhage), and general convulsions, especially on the same side as the lesion. The slower the hemorrhage the greater the chance of making a regional diagnosis. However, general convulsions usually supervene on account of the increased cerebral pressure. In infratentorial hemorrhage, the children are usually quiet, their symptoms are those of disturbance of the respiration, such as great irregularity and apnea. Lumbar puncture fluid is often bloody; there are no unilateral symptoms, the convulsions are usually bilateral, and tonic convulsions may predominate; opisthotonus, retraction of the head, and erection of the penis are often present. The cortical symptoms are late, due to edema of the brain, and manifest themselves in ocular motor symptoms, tonic and clonic convulsions.

Beneke who has studied hemorrhage over the base of the occipital lobes, showed that they had their origin in a characteristic tearing of the tentorium. Through compression of the skull the falx cerebri is lengthened and tears; the tear extends into the tentorium in which some fibers from the falx terminate.

Hemorrhages into the brain substance are usually due to very severe births. They are followed by general convulsions; there is no blood in the cerebrospinal fluid. The symptoms may come on late, often after three or four days. Hemorrhage and resulting convulsions may be due to an underlying hemorrhage diathesis especially in the case of premature infant—brain trauma without hemorrhage is becoming more recognized as a cause of convulsions and death in the newborn.

CONVULSIONS IN INFANTS AND CHILDREN (FROM THE END OF THE NEWBORN PERIOD TO THE THIRD YEAR)

After the newborn period is passed the frequency of convulsions diminishes and the various causes do not seem potent during the next few months of life. From the sixth, or better, from the eighth month, convulsions again become more prevalent and continue so until between the second and third years. The occurrence of convulsions at this period is ascribed by many to the rapid growth of the brain and the spinal cord and to the fact that the adult inhibitory factors in the cerebrospinal system are not yet formed. At birth the average weight of the brain is 370 gm.; the adult brain-weighs 1250 gm. Thus the brain adds about 1000 gm. to its birth weight. Three hundred of these are gained in the first nine months, at the end of which the brain weighs 670 gm. At three years, the weight has reached 970 gm., three times its birth-weight. The medullary sheaths are being rapidly formed during the first year of life. The projection system is only completed by the ninth month; the tangenital fibers as well as the association fibers are formed about the same time only after years; the former, however, are present in small parts at birth.

The fact however, that convulsions are relatively uncommon from the second to the sixth or eighth month leads one to the conclusion that this is due not to the rapid development of the central nervous system but rather to the absence of rachitis, tetany, etc. at this period.

Convulsions at this age may be broadly divided into: (1) Functional, including toxic, and (2) organic, including bacterial infections. We will concern ourselves mostly with the purely functional group and take up the toxic, bacterial, and organic causes when discussing the convulsions occurring during childhood and puberty, for the etiologic factors are much the same. The great group of convulsions in the first three years of life without any pathologic change in the central nervous system, classes collectively as, *Eclampsia Infantum*, occurs in many children: They are often repeated and are both clonic and tonic in character. There is frequently no demonstrable lesion which would at any age produce a convulsion. The children may be perfectly well or more or less severely rachitic. These convulsions were considered by Soltman as physiologic, due to a rapidly growing nervous system, in which as the inhibition was still imperfect, an irritant from the periphery (Eclampsia Reflectorial) or a poison from an unknown cause (Eclampsia Idiopathica) produced a convulsive attack. This reasoning was shown to be wrong.(Bechterew). Escherich was one of the first to point out that many of these children had also suffered from laryngospasm, and Loos, and later Thimisch and Mann, were able to show that there was a change from the normal electrical (galvanic) response in these children; one that resembled the reaction that Erb found in adult cases of tetany. This consisted, as is now well known, of (1) an increased galvanic irritability of the peripheral nerves, so that less current is required to elicit a contraction than in the normal child; and (2) the open and closing contraction of the galvanic current differs from the normal. This is best shown by the comparison of the normal contractions with the reaction in tetany.

	C.C.C	A.C.C.	A.O.C.	C.O.C.
Normal	1.4	2.24	3.6	8.2
Tetany	0.6–0.7	1.0–1.4	0.5–0.9	1.9

From the above it will be seen that the responses to all currents are with less milliamperes in the case of tetany. Especially to be remembered is the fact that normally there is no response to the cathodal opening contraction until at least five milliamperes of current is used, whereas in tetany a contraction to the C.O. current is obtained with much less than five milliamperes.

This spasmophilic or latent tetany was supposed by Escherisch to be due to a disturbance in the secretion of the parathyroid bodies and many animal experiments and pathologic findings (Yanase) tend to support this. Schmorl, however, found the parathyroid bodies perfectly normal in a marked case of tetany. This does not disprove the above findings, as it is well known that a normal histologic finding does not preclude an abnormal functioning of a gland. There is certainly an abnormal chemical change in the nervous system as shown by the diminished amount of calcium in the brain (Quest) and blood (Howland and Mariott) of cases of manifest tetany. That a disordered metabolism is at fault was well shown many years ago by Flesch and also by Bohn, who put these children on breast milk and caused the laryngospasm to disappear. Fischbein eliminated cow's milk, gave a carbohydrate diet and cured the convulsions; and finally Gregor and Finkelstein showed that the electrical excitability could be changed by a change in diet. Lately Watanebe has shown that the injection of Quanidine Hydroch gave the same picture as tetany. This spasmophilic condition is rarely discoverable before the second month and usually not until the sixth month or more. It is extremely common. For a description of the clinical picture and treatment of the spasmophilic condition and of tetany the article on that subject must be consulted.

Reflex Causes of Convulsions.—That trauma, heat, overloading of stomach, colic, meteorismus, worms, renal calculus, phimoses, hernia, adenoids, foreign bodies in ear or nose, teething and so forth, can cause convulsions is doubted by many. Heat stroke as an altered condition of the nervous system may.

The other causes of convulsions at this age, to be considered later, are interesting in the light of the above facts, inasmuch as probably less of the toxic substance, less trauma, etc., is needed to bring on the outbreak in a spasmophilic child than in one not so affected.

CONVULSIONS IN CHILDREN (FROM THE THIRD YEAR TO THE AGE OF PUBERTY)

The general causes to be considered are:

(*A*) Functional
  1. Tetany
  2. Epilepsy
(*B*) Toxic
  1. Organic such as those secondary to infections and other general diseases, especially kidney conditions.
  2. Mineral poisons, drugs, etc.
(*C*) Organic diseases of the central nervous system.
(*D*) Terminal.

(A) **Functional.**—**Tetany** becomes more and more infrequent after the third year of life; laryngospasm and general clonic convulsions are rare. The tonic spasm dominates the picture. Carpopedal spasm and bronchotetany are often the main features of the clinical picture. Infectious diseases, which are so common during this period, often cause general convulsions, due no doubt to the specific poison of the general disease, yet also because of the underlying spasmophilic diathesis. The Chvostek sign is usually present, the Trousseau less often, and Erb's phenomenon can usually be found if looked for.

Chronic tetany continuing from infancy, is not very common, but relapses of an infantile tetany, becoming evident at puberty must be thought of. The persistent form of tetany may be kept up by an intestinal derangement. One of the extremities is usually involved, or the face alone may be attacked, with the picture much like that of traumatic tetanus. The main features of tetany, Erb's phenomenon, Chvostek, etc., must naturally be present before a diagnosis can be made, and it must not be forgotten that both the tetany and another disease may be present in the same child.

Epilepsy.—Epilepsy comprises a second large group of cases having convulsions during this period of life. One must not forget that epilepsy can begin in early infancy, for acccording to Finkelstein, 7 per cent. of all convulsions in childhood are really the first sign of epilepsy. Others rate the proportion as high as 22 per cent.

According to Birke one must differentiate, first epilepsy with intervals—second epilepsy with continued attacks (Epilepsia continua). Epilepsy with intervals seemingly starts when the patient is well on in childhood, usually between the fifth and sixth years. Upon careful investigation of the previous history, however, it will often be found that the child had one or more convulsions during infancy or early childhood. In spasmophilia, the convulsions are usually frequently repeated, whereas in this interrupted form of epilepsy they occur in isolated attacks. The length of the interval varies according to surroundings and social conditions. Birke, in his table given below, showed that 30 per cent. of his cases were the interval kind:

Age of Commencement	Number of Cases
2–3 years	2
3–4 years	2
4–5 years	2
5–6 years	1
6–7 years	7
7–8 years	1
8–9 years	1
9–10 years	1
10–11 years	0
11–12 years	1

During the intervals Birke and Thiemisch have shown that irritability and mental inferiority are usually present. Irritability especially is an important symptom according to Aschaffenburg. The patients are moody, irritable, have attacks of rage, and suffer with pavor nocturnus. Yet they may be exceedingly intelligent and have no signs of stigmata.

Cases of Epilepsy without Intervals.—These may begin in infancy with general convulsions, frequently repeated, or they may begin gradually with petit mal, and a gradual increase in the size and severity of the attacks. Children thus affected may bring the disease into the world with them, in conjunction with marked idiocy, but this is not necessarily the case, for there are many such children having excellent intelligence. Cases of epilepsy which begin in later childhood, often start after a bad fall, an acute illness, with or without an encephalitis. Other individuals are apparently perfectly normal and the terrible disease springs out of a clear sky.

Such divisions are only of clinical value and serve to direct one's attention to the diagnosis; the attack, especially of grande mal, is not the beginning of the illness, as the body has long since been prepared for the outbreak. Temperature is usually absent. However, this is not necessarily the case, because just as in tetany where the secondary condition causing the lesion may produce the temperature, and the spasmophilic condition the convulsions, so with epilepsy; the secondary condition causes temperature and the underlying condition brings on the convulsions in an epileptic child. The general clinical features of an epileptic attack are too well known to be dilated upon. The aura, the suddenness of the attack, the general convulsions, preponderance of nocturnal attacks, usually absence of temperature, all lend themselves to the diagnosis. Convulsions, especially at night without any temperature in an older child, should always be looked upon with suspicion.

Epilepsia Secousses.—Without the loss of consciousness, local twitchings may be the beginning of grande mal. Attacks of tic or lightning like contractions with the child bending backward and forward almost as if salaaming, is often the forerunner of haut mal.

Eclampsia Nutans.—This is a condition in which children have attacks of salaaming. This must be differentiated from hysteria; it usually occurs in idiotic children and is part of the general epileptic state.

Epilepsia Procursiva.—In which there is forced running wildly about—consciousness is not disturbed—usually ends in a real attack of epilepsy.

A combination of epilepsy and tetany has been remarked upon.

(B) **Convulsions due to toxic causes** including the infectious diseases.

In Measles.—Convulsions may usher in the disease. They may however, occur later during the first week; then they usually denote the onset of some complication, especially pneumonia.

In Scarlet Fever.—Convulsions occurring at the beginning of the disease are not nephritic in origin, but due to the toxin of the scarlet fever. Occurrence in the second and third weeks may be due to nephritis; they are usually preceded by apathy and somnolence.

In Typhoid.—They occur in the second and third weeks.

In Severe Malaria.—They may be the equivalent of a chill, usually accompanied with gastric symptoms and high temperature. The younger the child the more frequent and violent these convulsions.

In dysentery and diphtheria convulsions are rather uncommon.

In Pertussis.—They are very common and may be due, first to a hemorrhage in the brain, or secondly to the toxin of the disease. Whether the characteristic laryngospasm and its accompaniment of cyanosis can cause a convulsion is an open question.

Parotitis epidemica and varicella have rarely been accompanied by convulsions.

In Uremia.—Either twitchings or general convulsions may occur. The diagnosis naturally rests upon the diagnosis of the disease and should be treated accordingly.

Intestinal.—Intestinal disturbance as a cause of convulsions may be discussed in three groups; first, relative or absolute over-feeding with perfectly fresh and wholesome food; second, the ingestion of decomposed or spoiled food; and third, the decomposition of good food in an intestinal tract where there is faulty metabolism or incomplete absorption. To what extent the first condition produces convulsions is an open question. The ingestion of spoiled food as a cause of convulsions is also questionable, for very few of the substances isolated from decomposed food are toxic. Vaughin isolated a toxic protein from ice-cream which proved exceedingly poisonous; and in the last instance, the decomposition of food in the gastro-intestinal tract, principally the proteins and the end products, is such a clouded field, so difficult to understand, that one cannot definitely state to what extent such decomposition may be the cause of general convulsions. It is easily conceivable that any of these causes may act upon a spasmophilic child, or that some of the poisonous protein-split products may be an etiologic factor. It has been so long the custom to regard these as a dominating cause, that emptying of the stomach and the intestines has always been a prime factor in the treatment of convulsions in infancy and childhood. In most instances especially if temperature be present, the convulsions are not intestinal in origin but there is an underlying infection present.

Drugs.—There are two distinct groups of drugs which produce convulsions. The one of which strychina is an excellent example, increases the reflex excitability of the spinal cord; the convulsion originates in a sensory impulse or impulses which are carried to the spinal cord and distributed over the entire cord, thus producing contractions. of the entire musculature. These convulsions may be so rapid that they soon cause the various muscles to be in tetanic contraction In the case of strychnina the normal reflexes are at first exaggerated, the child becomes very restless, tonic contractions and finally tetanus occur. Caffein is another example of this group. The second group in contradistinction to the former starts in the higher centers and therefore has an epileptiform or clonic character rather than a tonic character. *Atropin* is such a drug and is very apt to produce convulsions especially in young infants. Cocain and santonin also belong to this group.

Lead.—Poisoning with lead is probably more common in children than generally supposed. Blackfan gives an excellent resumé of the subject of lead poisoning with special reference to the convulsions produced by lead. Sucking the hands and fingers and eating the enamel of iron beds and licking painted walls are some of the ways by which the lead is ingested. The usual signs of lead poisoning may be present; on the other hand convulsions may be the only symptom. These are clonic and tonic but are characterized by their persistence. Chloroform has to be resorted to in order to quiet them, and when this is discontinued the convulsions often start again. Recurrences are the rule. Lead in the urine and feces and stippling of the red blood cells are the diagnostic features.

(C) **Organic diseases of the central nervous system** such as meningitis, encephalitis, brain tumor, tubercle, etc. may cause convulsions during the course of the illness either directly or by indirect pressure. For a general discussion of these conditions, the special chapters must be consulted.

(D) **Terminal Convulsion.**—Convulsions just before death are a common occurrence in infants and children, but especially so in cases of gastro-enteritis and heart disease. Whether these are due to the toxin of the underlying disease or to the disturbance of the circulation usually present is still an open question.

Differential Diagnosis.—(1) *Careful History:*

2. *Age of Child:*
   In the newborn more likely to be organic.
   Under three months—rarely tetany or spasmophilia.
   Six months to three years—look for spasmophilia.
   Erb and Chvostek signs.
   Do not forget that spasmophilia may be incidental and an organic disease also be present.
   Three years and over—look for infectious disease; if repeated over 24 hours look for organic disease of the nervous system and do lumbar puncture.
3. *With Temperature:*
   (*a*) Infectious disease.
   (*b*) Acute disease of nervous system; if repeated and lasting more than 24 hours do lumbar puncture.
   (*c*) Retraction of head more likely in organic cases.
   (*d*) Beginning locally and remaining so for some hours, more likely organic in origin.
   (*e*) Markedly accentuated orbicularis contractions with very little or no involvement of the extremities—usually denote organic origin.
   (*f*) Gastro-enteric in origin.
4. *Without Temperature:*
   Spasmophilia—test galvanic reaction.
   Epilepsy—convulsions occurring in older children without temperature with previous complete well-being, followed by some drowsiness, and quick return of normal condition.

Prognosis.—This depends upon a careful examination of the child from head to foot including the ears, eyes and the urine. The age of the child is an important factor. Between the ages of six months and two years functional causes are the most likely and after three years with temperature may be the precursor of an acute illness. Convulsions in older children without temperature should always be regarded suspiciously; repeated attacks with short or longer intervals of days or still longer intervals, should always make one think of epilepsy.

Treatment.—Immediate treatment should be directed toward elimination of any retained intestinal contents; an enema is given at once—stomach washing except in a suspicious history of acute food, metallic, or drug posioning need not be done. A warm bath with or without mustard is then given, but the parents must be warned that this bath need not be extremely hot for a very common cause of generalized scalding in infants is due to immersing in boiling water to allay convulsions. Should the convulsion not stop then a mixture of chloral hydrate and sodium bromid in a little water is given per rectum. The dose for an infant of one year is Chlorate Hydrate gr. v, Sodium Bromid gr. xv in 1 oz. of water. If the chloral is expelled or the convulsion continues morphine may be administered hypodermatically in doses of 1/50 of a grain for a normal size six months old infant, 1/30 of a grain for a normal size year old child. At the same time chloroform may be administered in doses sufficient to control the convulsive movements but no more. If the child continues to have convulsions over many hours lumbar puncture may be performed as a theraputic measure, although we have not seen much benefit from it. It is a good rule, however, especially in infants, that when convulsions are repeated for more than 24 hours lumbar puncture should be done for diagnostic reasons if for no other. Naturally a careful search must be made for the underlying cause which must be attacked.

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CHAPTER CLXXIII

NEUROSES

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EPILEPSY

Definition.—Epilepsy is a symptom-complex characterized by a number of morbid phenomena, of which the most striking and common are periodic attacks of loss of consciousness, convulsions, and so-called post-epileptic states. Such phenomena are known as major epilepsy (grand mal). In a very large group of cases the clinical picture is incomplete, some of the cardinal symptoms only being manifest. For instance, the entire epileptic attack may be confined to a transient loss of consciousness, to an abortive convulsion, or to so-called premonitory symptoms. Such cases are described as mild or minor epilepsy (epilepsia mitior, petit mal). Besides the major and minor epilepsies, there is a third group in which the predominant features are purely motor or sensory-motor, confined to some portion of the body, to one limb or to a part of it. This type of epilepsy usually indicates an organic lesion of the brain, preferably its cortex, and is described as organic, symptomatic, cortical, partial or Jacksonian epilepsy. The latter may be more properly designated as "epileptiform," as contrasted with "epileptic" states which are specific for the grand and minor attacks, and which are also known as genuine *idiopathic*, or *generalized epilepsies*.

In addition to the foregoing types, such conditions as eclampsia ("acute epilepsy"), reflex epilepsies, tuberous sclerosis epilepsy (Bourneville's disease), myoclonus epilepsy and continuous epilepsy (Koschewnikow's disease) are described. Epilepsies are also classified according to their etiology. If caused by trauma, they are called traumatic; if caused by syphilis, syphilitic; if by cerebral hemiplegia hemiplegic, and so forth. It is evident that the latter types should rather be designated as epileptiform states or symptomatic epilepsies since in them it is possible to demonstrate some organic brain lesion which causes the epileptic manifestations. The symptomatic epilepsies have nothing to do with epilepsy proper, in which no typical gross or microscopic brain lesions or a dependable etiology can be demonstrated. Epilepsy is a chronic disease process, wholly made up of paroxysmal attacks of unconsciousness, convulsions, and many other more or less inconstant features, while an epileptiform state is merely a symptom of some other morbid condition. Some authors, as Pierre Marie,[1] Lemoine,[2] Redlich,[3] Souques,[4] Claude and many others, consider genuine epilepsy as a mere symptom. In other words, according to these writers, epilepsy is symptomatic, signifying some macroscopic or microscopic cerebral lesion. However, since no definite pathology of the so-called, idiopathic epilepsy has been established, it would be proper to adhere to the old classification of Jackson: epilepsies (major and minor) and epileptiform states.

Symptomatology.—Major epilepsy is characterized by the occurrence of so-called major or grand epileptic attacks (seizures, fits). In its typical complete form, a major epileptic attack exhibits several features: First, premonitory symptoms (preludes) or prodromes; second, aura; third, loss of consciousness, preceded by an epileptic cry and followed by tonic and clonic muscular contractions; fourth, post-epileptic states (stupor, sleep, loss of sphincter control, amnesia, that is, loss of memory as to occurrence of the attacks). The premonitory or prodromal symptoms more or less remotely precede the attack. They show as motor, sensory, vasomotor, visceral and psychic disorders. Thus, long before the attack sets in, hours or even days, the patient may experience slight jerking in various parts of the body, or a mere tremor or muscular weakness (*motor* prodromes); or there may be *sensory* manifestations—paresthesias, pain, ringing in the ears, dizziness, photophobia; *vasomotor* disturbances—eruptions on the skin in the form of erythema, urticaria, changes of the skin color; *visceral*—gastric or intestinal disturbances (pain, nausea, vomiting, diarrhea), and *psychic* symptoms. Changes in patient's conduct, character, disposition, behavior and speech are noticed by friends and relatives. These patients may be either depressed or excited, overactive, quarrelsome, and do not sleep well or do not sleep at all.

In some cases, the premonitory symptoms are absent, while in others *they may constitute the entire attack;* that is, in some cases of epilepsy, there is no aura, nor convulsion. For a long time the true nature of the various vague complaints, which may pertain to the motor, sensory, psychomotor spheres, remains unrecognized. Later they are followed by definite symptoms (aura, convulsions, coma, and then it becomes apparent that the foregoing periodic complaints were abortive or mild attacks of epilepsy.

The prodromal stage is followed by an *aura* which usually merges into it, and which differs from it only by its short duration. The aura lasts a few seconds, sometimes sufficiently long to enable the patient to take the necessary precautions to meet the attack. (Children usually place themselves on the floor.) Like the premonitory symptoms, the aura may be motor, sensory, visceral, vasomotor or psychic. Thus, tingling, pain and needle sensations, numbness in the limbs, tongue, face, visual and auditory sensations (colored vision, whistling, roaring), epigastric pains with a sensation of a ball ascending to the throat from the stomach or intestines; palpitation of the heart, occur. In short, any organ of the body may participate in originating an aura, or may be the starting point of an epileptic attack. Especially frequent and important is the psychic aura, in which the patient performs strange acts, such as rubbing his forehead, buttoning and unbuttoning his clothes, or undressing himself; or when he may show purely psychic disorders, such as fears, depressed states, hallucinations and illusions. Like the prodromes, the aura is sometimes the sole manifestation of an epileptic attack and may be confined to any of its numerous symptoms. For instance, a patient may complain of vague gastric disorders, such as nausea, pain, faintness in the stomach, such complaints occurring periodically, in the same manner and intensity, for months and years, coming in paroxysms, i.e., with free intervals, until followed by a typical epileptic fit. Similar paroxysms may occur as intestinal, cardiac, pulmonary or laryngeal disorders, diagnosed and treated as such, while in reality they are but an aura, or an abortive epilepsy (Herpin).[5] On the other hand, in about 50 per cent. of cases Féré[6] and Gowers[7] say that the aura is absent, or, what is probably more correct, so short that it is unnoticed.

In typical cases, the aura is followed by loss of consciousness, initiated by a cry, sudden fall and convulsions. The epileptic cry is rather a scream, resembling that of a person being strangulated. It is loud, a cry of agony (Herpin), being caused by sudden tonic contractions of the muscles of the throat and expulsion of the air through the larynx. The patient falls to the ground, irrespective of the place and surroundings. When the aura is absent, the fall is sudden, owing to the loss of consciousness, which is always complete and synchronous with the convulsions. The latter are tonic and clonic, usually affecting both sides of the body, but they may also be unilateral.

In the tonic stage the head and eyes are turned to one side. The arm is flexed to a right angle at the elbow joint and slightly abducted at the shoulder joint. The wrist and metacarpophalangeal joints of the fingers are strongly flexed. The terminal phalanges extended, the thumb strongly adducted, the lower extremities extended, with the feet inverted. The tonic stage lasts but a few seconds, being followed by the clonic phase, in which the muscles of the face, tongue, throat, larynx and of mastication are especially involved. The tongue is often caught between the teeth and bitten. Frothing at the mouth occurs, and at times may be bloody. The head is banged against the floor; the eyes are rolled sidewise; the face is cyanotic and livid. The pupils, according to Binswanger, are contracted during the initial cry, dilated in the tonic state and again contracted in the clonic, and they do not react to light, while the conjunctiva and cornea, as well as the entire body, are devoid of any sensation. The tendon, skin and other reflexes are lost. Incontinence of the bladder and frequently the rectum is manifested, and often there are seminal emissions. The convulsive stage lasts from one to three minutes, followed by stupor or epileptic coma, and sleep, from which the patient wakes without recollection or knowledge of the attack.

Cases of epilepsy occur in which either the entire convulsive stage, or some of its phases, such as the initial cry, the tonic or clonic spasms, or the involuntary bladder and rectal discharge are absent.

In some instances the attack may be confined to convulsions only, without prodromal symptoms, aura or fall, or it may be represented by a transient loss of consciousness (the classical petit mal), and, according to some authors, by periodic attacks of involuntary urination. Herpin describes cases in which the entire epileptic attack was represented only by the initial cry.

The after attack, or post-epileptic state, usually consists of a prolonged sleep, of two or more hours. When the patient wakes, he usually has an intense headache, weakness, exhaustion, soreness in the body, tongue and mouth. These symptoms, as a rule, soon pass away, but there may be other post-epileptic signs, such as hemorrhages in the conjunctiva, cornea and skin, which persist much longer. The patient usually recovers rapidly from the attack. In some cases, muscular weakness remains, in the form of paralysis of one-half of the body, including the face and tongue (hemiplegia), often associated with aphasia, apraxia, agraphia, visual disturbances (amaurosis) and contracted fields of vision. The post-epileptic paralyses, ascribed by some authors to the exhaustion, by other, to the inhibition of the cortical centers, may last hours, or days, or may remain stationary, especially after severe attacks and so-called epileptic states. Cases are on record in which the entire epileptic seizure was confined to imperious, sudden attacks of somnolence, or sleep, of several hours' duration. In Kesteven's case,[8] a child had periodic attacks of stupor, which were interpreted as abortive epilepsy and successfully yielded to treatment with bromids, while Jacob Weiss[9] speaks of an epileptic patient whose attacks sometimes consisted of a sleep of several hours' duration. Instead of sleeping attacks there may occur psychomotor disorders (equivalents). The patient suddenly runs for some distance, either in a straight line or in a circle; or he performs rotary movements, or he rolls on the floor. This is the so-called *procursive* epilepsy in which the motor phase of the attack, the convulsions, is replaced by running acts, the attack ceasing with the cessation of the running. In some instances, the running replaces the aura, and in exceptional cases it may form the post-epileptic phase (Semmola's case). Mairet[10] remarks that the running is swift, the victim either avoiding possible obstacles in his path or paying no attention to them, whatever they may be, fire, water, precipices, etc. After a certain distance has been covered, the patient stops, returning to the spot whence he started. The running, preceded by a series of cries, is followed by great fatigue and exhaustion, the patient having no knowledge of his strange spell. Ultimately, according to Mairet, the running or procursive epilepsy is transformed into an ordinary classical type, or the classical and procursive attacks alternate. The latter may also occur at night when the patient is asleep. He leaves his bed and starts running from one bed to another, or through the rooms, or rolls around or up and down his bed.

Finally, cases of epilepsy occur in which the entire attack is replaced by psychic disorders. The patient, instead of having convulsions, performs strange acts, such as undressing himself in a public place, exposing his genitals or undertakes purposeless, aimless wandering. He suddenly leaves his home, wanders around until found by strangers in some remote place. Such attacks of unconscious wandering, generally classified as *dreamy* states, fugues, pyromania, ambulatory automatism, are also known as epileptic equivalents, or psychic epilepsy (compare with chapter on hysteria).

Cases are on record in which patients have made prolonged journeys, transacted business and kept records of every occurrence without exciting any suspicion of being in an abnormal condition, and without recollecting any event after recovery. Such cases of psychic epilepsy, or equivalents, either replace the epileptic attack or precede it, forming its prodromal stage or aura. The psychic equivalents also appear as mental disorders, such as impulsive acts, unprovoked assaults, indecent performances, homicidal tendencies, stealing and lying, indecent performances, homicidal tendencies, stealing and lying. Criminal acts may be committed by the patient while he is unconscious. According to Spratling,[11] he knows nothing of his misconduct until he is informed, when he usually falls into great depression, or has a typical convulsion which breaks up the abnormal state.

The major attacks usually occur at night (nocturnal epilepsy). Minor epileptic attacks generally occur during the day (diurnal epilepsy). A sufferer from nocturnal epilepsy, if not watched during his night attacks, may not be aware of the malady for many years, as upon awakening he does not recall that he had an attack while asleep.

The characteristic feature of epileptic seizures is their periodic, paroxysmal occurrence. In some cases there may be but one attack daily, weekly or monthly. In milder cases the intervals may be several months, while in severe cases the attacks may be in a series, that is a great number of attacks may occur during the same day, and after an interval of days or weeks another series of attacks may begin. If they follow one another without free intervals, that is, without the patient regaining consciousness, it is known as status epilepticus (état mal). It usually indicates some organic brain lesion, or in children, according to H. Vogt,[12] some disorder of metabolism.

2. Minor Epilepsy and Incomplete Attacks.—In its classical form, minor epilepsy is represented by slight attacks of unconsciousness, that is, sudden suspension of the mental functions. The patient suddenly turns pale, stares vacantly before him, and with equal suddenness recovers his normal mental state. Thus, he may suddenly stop in the middle of a word or a sentence, suspend writing or working, and with the return of consciousness, continue the interrupted function, without being aware of what occurred. Aura, cry, fall, convulsions and other phases of an ordinary major attack are absent, the only element being the unconsciousness. In short, minor epilepsy may be designated as a type of so-called *incomplete* epilepsy. As studied by

Herpin these are of great interest and practical value. While speaking of the various phases of a major epileptic attack I have already pointed out that it may be reduced to one of its numerous stages, such as, prodromes, aura, or post-epileptic state. As these stages comprise an enormous number of clinical manifestations (motor, sensory, visceral, psychical), it can easily be understood that so-called atypical, incomplete attacks may be of numerous varieties. For instance, the abnormal paroxysmal sensations (paresthesias) in the fingers, hands and toigue may, for a long time, be the sole manifestations of epilepsy. Gastric disorders (abdominal pains, nausea, periodic vomiting, sense of pressure), intestinal disorders (abdominal pain, diarrhea), urinary disturbances (excessive desire to urinate), abnormal sensations in the pharynx and larynx (suffocation sensation), in short, disturbances of any organ of the body may form not only the starting point of an epileptic attack, that is, begin the latter, but may be its sole manifestation. Such vague visceral or other sensory disturbances may, according to Herpin, persist for years, occurring in paroxysms, i.e., with free intervals, and always showing the same clinical features, until followed by definite manifestations of major epilepsy. And only then is the true nature of the foregoing visceral disturbances and other indefinite complaints properly diagnosed and treated.

One remarkable type of an incomplete epilepsy pointed out by Herpin deserves special attention, so-called *epileptic commotions.* These are sudden jerkings involving either the entire body or one limb or a portion of the latter (hand, finger, foot, toes, etc.). The commotions last but a moment, and impress one with their suddenness; the patient having no time to take the necessary precautions. During such a commotion, the hands, for instance, suddenly relax and drop their contents. In some cases, the sudden jerkings are combined with upward turning of the eyes, or with temporary loss of eyesight, or even slight loss of consciousness. The further course of such commotions is either unaltered, for as long as 15 years, or may become complicated by typical epileptic attacks.

Whatever the type of the incomplete attacks, they are exceedingly frequent, much more frequent than the classical minor or major fits. Herpin asserts that only 5 per cent. of the epileptics do not show these incomplete features and if they are looked for they can be detected long before the regular typical attacks occur. By the time these do take place, he says, the disease is already deeply rooted, making the prognosis much worse than when the trouble is recognized earlier. It should be noted that many types of epilepsy referred to by Herpin as incomplete, with their origin in the peripheral nerves, are undoubtedly so-called Jacksonian epilepsies, or epileptiform states, and, therefore, do not belong to epilepsy proper.

The clinical picture of epilepsy, however, is not confined to attacks only, whether complete or incomplete. Even in the so-called intervals free from attacks, a pronounced epileptic exhibits features which stamp him as a pathologic individual. These features may be *physical* and *psychic.* To the former belong so-called stigmas of degeneration, shown by the shape of the skull, ears, teeth, face, and voice. The face, for instance, in many cases is plump and rough, with thickened, "sausage"-like lips and a disagreeable expression; the voice, studied by Clark and Scripture,[13] and L. P. Clark[14] is harsh, and monotonous. But of greater importance and significance are the psychic symptoms which may be apparent long before the physical symptoms become evident. A potential epileptic, says Clark,[15] shows egocentricity, supersensitiveness, marked emotional poverty and rigidity, lability of mood—traits which defy all ordinary systems of training. In school many of them do not concentrate; they do not adapt themselves to the school routine and are restless and irritable. In their actions and play they are slow and clumsy; many are disobedient. The intellect is not so much at fault as the emotional reactions to the social environment. Many epileptics show great fatigue, are irritable, restless, depressed or impulsive. Ethical defects (morbid lying, inclination to steal) are typical in the so-called epileptic character. By these psychic features epilepsy is stamped not only as a nerve disorder, but also as a mental malady, placing this disease on the borderland of psychic disorders.

3. Eclampsia Infantum.—Infantile eclampsia, acute epilepsy of Hasse, Féré and other older authors.

Infantile eclampsia is a type of convulsion occurring in very young children. Their age is not below 2 months and not above 18 or 20 months, though, according to Thiemich[16] and Birk,[17] the attacks may recur at the age of 8 or 10 years (delayed eclampsia), when they resemble major epilepsy. They are especially frequent during the first six months of the child's life, eclampsia representing the most frequent type of convulsion in children in general. D'Espine[18] calculated that out of 100 children suffering from convulsions 61 have eclampsia, 33 have laryngospasm and 6 tetany. These three types of convulsive states make up what the German authors (Finkelstein, Thiemich, Birk) call the spasmophilic diathesis. By this term they classify a constitutional anomaly affecting the motor system and manifesting itself in electrical and mechanical over-excitability of the nervous system. The cathodic opening contractures occur with weaker currents, and the anodal opening contractions take place before the anodal closure contractions, contrary to the normal formula. The mechanical nerve excitability (Chvostek and Trousseau signs) is also increased. The increased excitability of nerves and muscles creates a predisposition to partial or general clonic and tonic convulsions. Spasmophilia occurs at certain times of the year, at the end of winter and the beginning of spring and may be provoked by improper diet. Birk asserts that in his cases none of the spasmophilic children became epileptics, that spasmophilia and epilepsy are entirely different morbid entities. Other authors, such as Potpeschnigg,[19] Redlich[20] and Hochsinger, do not accept such teaching, declaring that spasmophilia and epilepsy may co-exist and that epileptics may be found among former spasmophilic infants. At any rate, observations have not been made for a sufficient period to warrant rejection or acceptance of either teaching. The fact is that many children in early infancy show epileptiform states which must be properly defined and differentiated from real epilepsies (discussed in the section "Differential Diagnosis").

In infantile eclampsia the convulsive attacks are usually accompanied by fever. As a rule, they terminate in complete recovery, except in a few obscure cases which subsequently become ordinary epilepsy. Some authors of the older school, such as Baumes and Féré, even go so far as to assert that there is no reason to differentiate between infantile eclampsia and epilepsy. In the light of the theory of spasmophilic diathesis, this view is decidedly wrong, at least in the great majority of cases. For instance, it can be shown that the mother of an eclamptic child herself suffered in her infancy from eclampsia without becoming an epileptic in later life.

The convulsions in eclampsia consist, as in epilepsy, of a tonic and clonic stage. They do not follow the typical cycle as in an ordinary epileptic attack but are rather irregular, and mixed. They are accompanied by loss of consciousness, cyanosis, and after a few minutes they terminate in profuse sweating, while the initial cry and the post-epileptic phase (stupor and sleep) are absent. In older children Thiemich described an aura—restlessness, absentmindedness, irritability, increased emotivity.

The convulsions may be either localized (around the mouth and eyes), or generalized. Beginning in the bulbi of the eyes, they spread over the face, neck, upper extremities, trunk and lower limbs, being, as a rule, bilateral and symmetrical. The attacks usually occur in series, in "batches" of from 10 to 20 in 24 hours. The intervals between the series may be days, weeks or months, depending on the cause and treatment.

Etiology.—The causes are predisposing or exciting. The former, the spasmophilic diathesis, have been discussed already. The exciting causes are digestive disturbances, as overloaded stomach, artificial feeding, constipation, febrile diseases and respiratory difficulties (excessive crying, coughing, etc.).

Differential Diagnosis.—Infantile eclampsia should be differentiated from brain lesions, such as meningitis or encephalitis, and especially from epilepsy, for an infantile convulsion may signify either epilepsy or eclampsia. The latter is not observed before the eighth week, and only exceptionally after the twentieth month (delayed eclampsia), while epilepsy does not spare any period of life however early; eclamptic convulsions, as a rule, occur in series, epileptic convulsions, as single fits. In breast-fed children eclampsia is rare, while epilepsy may occur in any child, regardless of the mode of feeding. In eclampsia, spasmophilic signs can be discovered; in epileptics, they are absent. The former occurs preferably at certain times of the year; the latter irrespective of seasons. Some causes, such as faulty diet, can be discovered in eclampsia, none in epilepsy; in epilepsy there may be defective heredity; in eclampsia a history of eclampsia in the mother. Free periods occur between the attacks in eclampsia, while in epilepsy there are practically no completely free intervals. For instance, the child shows irritability, suffers from bed wetting, or night terrors, which disorders ultimately may turn into a typical epileptic fit. According to some authors (Birk), if convulsions do not cease after proper dietetic measures, or if they cannot be accounted for, epilepsy should be suspected. Patrick and Levy [Early Convulsions in Epileptics and in others, J. A. M. A., lxxxii (1924), p. 375] studied the significance of early convulsions in a group of 500 epileptics in private practice and compared them with a group of 752 unselected infants and children. In the epileptic group the frequency of infantile convulsions was five times as great as in the unselected group. The true interval between infantile convulsions and epilepsy varied from several months to 35 years. The possibility of an early convulsion being a forerunner of epilepsy is greater if it occurs before 6 months or after 17 months than if it falls within these limits. Convulsions over a long period of time (three hours) or followed by confusion or torpor, or limited to one limb or one side of the body, are noted exclusively in the epileptic series, the non-epileptic convulsions being typically brief, generalized and not followed by confusion or prolonged stupor. "Teething spasms" occurring between 11 and 13 months appear to be especially benign.

The foregoing principles, in the majority of cases, help in establishing the true nature of epileptiform states in young children, though some need still further elaboration. The prognosis of eclampsia is good—complete recovery being the rule.

Treatment.—Eclampsia demands dietetic and hygienic treatment principally, that is, proper feeding, proper regulation of the bowels, fresh air and plenty of sunshine combined with cod-liver oil—in short, the same treatment as that used in tetany and other spasmophilic states.

4. **Reflex epilepsies** are produced by some peripheral nerve lesions or diseases of any organ in the body (skin, teeth, nose, throat, ears, intestines, or genito-urinary organs). Féré calls such localities epileptogenetic zones, as their irritation may result in an epileptic fit. Carious teeth, foreign bodies in the ears, eye, or nose, polyps in the nasal cavities, diseases of the pleural cavities (empyema) are occasional causes of epileptic attacks. All these external factors may be instrumental in causing epilepsy, provided there is some predisposition to this malady. Of especial interest are the reflex epilepsies caused by intestinal worms (*Oxyuris vermicularis*, pin worms). Some reliable reports in the literature, such as Martha,[21] for instance, speak decidedly in favor of such an etiology (verminiform epilepsy of the French). The attacks of verminiform epilepsy may occur at any age (from 3 to 45 years), in varying intervals. They have some of the features of an ordinary epilepsy, such as aura, loss of consciousness, post-epileptic states, but differ in the absence of the initial cry, pupillary changes, fall and tongue biting. The aura and post-epileptic stage are much prolonged, while the convulsions, tonic and clonic, as well as the coma,

last much longer than in an ordinary epilepsy. With the expulsion of the parasite, the epileptic attacks cease. The reflex epilepsies rarely occur, and ought not to be designated as epilepsies at all, but rather as epileptiform states. They are included in one group together with minor epilepsies and eclampsia, because, though possessing a definite etiology, they, like the previous three conditions, show no definite pathologic changes.

Before we proceed with the discussion of epileptiform states, the differential diagnosis, pathology, prognosis and treatment of epilepsy proper will be considered.

Differential diagnosis of epilepsy is to be made relative to the attack itself, in its typical form, and its various components (premonitory stage, aura, convulsions, post-epileptic state).

The first question in every case is to determine whether we have to deal with a case of true epilepsy or an epileptiform state, i.e., whether the condition is merely a symptom of some brain or constitutional disease. The best means of arriving at a diagnosis is for the physician to witness the attack, though in some cases the description and history suffice to show the true nature of the attack. When the latter, however, is confined only to the aura, or to the slight loss of consciousness, to the premonitory or the post-epileptic stage, a differential diagnosis is very difficult. As a rule, a single symptom should not be relied upon for diagnosis, but one should rather group the symptoms in conjunction with other events, such as the history, course and serologic findings. For instance, twitchings, jerkings in the limbs or muscles of the face may be due to epilepsy, but they also occur in chorea, tics, hysteria, spasms, and Jacksonian fits, from which they can be differentiated by considering other features peculiar to these morbid states. The same is true for the aura, which may be identical in hysteria, epilepsy or migraine. The numerous motor, sensory, visceral and psychic symptoms of the premonitory stage and the aura which, as noted, constitute the incomplete epileptic attacks of Herpin, should always be thought of as possible epilepsies. Herpin pointed out that though the nature of such incomplete attacks may vary *ad infinitum*, they almost always are identical in the same epileptic, always recurring in the same manner, running the same paroxysmal course, ultimately resulting in a typical epileptic fit of which the vague visceral, motor, sensory and psychic symptoms were the initial stage (preludes). The paroxysmal feature of the various morbid states, the comparatively good condition of the patient in the intervals, the negative findings of any organic or functional process that might account for the vague morbid conditions, help in the proper diagnosis of epilepsy. To the category of obscure and incomplete types of epilepsy proper probably belong some instances of so-called "vagal" attacks. The latter are defined by Gowers[22] as sensory symptoms "comprehending subjective gastric, respiratory and cardiac discomfort, sometimes cardiac pain and a sense of impending death, a sense of vague fear and dread." The attacks seldom last less than 10 minutes and may continue for a half hour or more. The onset is sudden, the ending gradual, the seizures recurring at varying intervals, accompanied by faint pulse, pallor of the face and slow cerebration. Sometimes they interpose in the course of a regular epileptic attack, as if, says Gowers, "symptoms usually momentary, as the aura of epilepsy become protracted, extended, as it were, into a vagal attack." Gowers could stop such attacks with nitroglycerin, indicating a disturbed function of the vagus nerve, and he thinks they are of epileptic character, or rather belong to the borderland of epilepsy. It should be pointed out that vagal attacks sometimes also indicate a gross cerebral lesion, such as tumors of the acoustic nerve and hydrocephalus, from which their epileptic nature should be differentiated.

The epileptic attack itself, with loss of consciousness, cyanosis, foaming, biting of the tongue, tonic and clonic convulsions, post-epileptic states, amnesia, is usually sufficiently characteristic to differentiate it from an hysterical attack. In the latter the consciousness is usually retained; the cyanosis is absent; foaming and biting of the tongue are never present; the convulsions are of the tonic type; the post-epileptic coma and total amnesia are absent. The initial cry is also lacking, as well as the indiscriminate fall associated with the unavoidable injuries of the epileptic. If an hysteric patient falls, he usually chooses a proper spot; the fall is deliberate. On the other hand, the grand movements of an hysterical attack described in the proper chapter, as well as other features, are absent in epilepsy. The post-epileptic conditions, such as dreamy states, ambulatory automatism, insanities, speak rather for epilepsy than hysteria.

In petit mal attacks when the loss of consciousness is transient and the patient grows pale, an ordinary fainting or syncope should be considered. In syncope the heart action, as well as the pulse, is faint, and weak; the face at the beginning of the attack is pale and covered with sweat; there is great weakness and interrupted breathing. In petit mal, the heart action and pulse are rather strong, the patient does not gradually sink to the floor as in fainting; after-effects, such as weakness and exhaustion, are absent, while, according to Gowers, the pallor *follows* the onset of the attack. Petit mal patients may show physical signs of degeneracy or intellectual defects, or they may be seized with a grand mal attack, which makes the diagnosis clear. The coma and post-epileptic sleep can be diagnosed from the history, condition of the reflexes, temperature, and, what is most important, from the subsequent course. A history of epileptic seizure preceding the coma would speak for epilepsy. The presence of Babinski's sign and exaggerated reflexes also speaks for epilepsy, while the enormal temperature, the condition of the urine and the blood pressure may help to diagnose the condition from other forms of coma, caused, for instance, by uremia, diabetes, or cerebral hemorrhage. Psychic equivalents without proper history cannot be differentiated from other forms of acute insanities, such as exhaustion or confusional psychoses. Malingering or feigned epilepsy differs from true epilepsy by the presence of all the reflexes, including conjunctival and corneal, and especially by that of the pupillary reflex which cannot be simulated.

Etiology.—The actual cause of epilepsy is not known. The prevalent opinion is that a defective heredity—direct or indirect—is its underlying cause. In direct heredity, one or both parents suffer from epilepsy; in indirect, the parents or other near relatives suffer from some other form of constitutional or mental disorder, such as insanity, diabetes, alcoholism, syphilis or tuberculosis, which, as H. Vogt remarks, reduced the vitality of the offspring, predisposing it to epilepsy. In general, Finkh[23] says, in about 50 or even 70 per cent. of the cases, a psychoneuropathic or some other defective heredity can be detected. In the United States and Europe, according to Gowers[7] and Turner,[24] about three persons out of every thousand inhabitants suffer from epilepsy. In more than 78 per cent. of cases, epilepsy commences between birth and the age of 20. Of this amount, according to Gowers, about one-third occur in the first decade of the patient's life; 46 per cent. in the second decade, about 6 per cent. in the third and fourth decades. According to Spratling's statistics, 13.5 per cent. begin during the first three years of life. However, it is not always possible to determine the exact time of the onset of the disease, as the attacks often occur at night, and for a long time escape attention. Turner and Spratling's statistics show that during the first year of life the onset is most common; the number of cases falls rapidly between the first and fourth years; from the latter age there is a slight increase to the seventh year. The second maximun is reached at the ages of 12 to 15, preceded by a decline at the ages of eight and nine. In other words, the first years of life and the time of puberty are the commonest periods of onset.

In the presence of faulty heredity, numerous exciting causes are instrumental in causing epilepsy. In this class belong the infectious diseases, head injuries in early childhood, intoxications (lead), physiologic factors (periods of puberty and pregnancy), and emotional shocks (fright, for instance). Some authors, for instance Lemoine and Marie, consider the foregoing factors, especially infectious diseases and intoxications, of prime importance in the causation of epilepsy. If the child was born an epileptic, as they admit may happen, then the noxious causative factors—infection and intoxication—probably affected the victim during its intra-uterine life. Marie could find, in every epileptic, a history of attacks dating back to the earliest infancy, and he thinks that however deficient or faulty the heredity might be, in the absence of an infection, there can be no epilepsy. Whether this theory of Marie or the usually accepted teaching of defective heredity is correct is impossible to tell, so long as our knowledge of pathology is defective. Only the exact pathology, if there is such, can tell us what is the main cause of epilepsy.

Histopathology and Physiology.—The histopathologic changes in the brains of epileptics, as described, are not sufficiently uniform and constant to justify the speaking of a pathology of epilepsy. The subpial gliosis of Chaslin,[25] the unilateral or bilateral sclerosis of Ammon's horn, capillary hemorrhages, atrophy of some portions of the cortex, swelling and atrophy of ganglion cells, proliferation of the neuroglia over the entire cortex following the atrophy of the ganglion cells (in the second and third cortical layers) and nerve fibers, misplaced and dislocated nuclei, hyperemic vessels, slightly infiltrated and thickened meninges—all these changes do not signify anything specific, and do not permit a pathologic diagnosis of epilepsy. They are inconstant, being more or less pronounced only in old cases, especially when complicated with dementia. In many cases Alzheimer[26] found no changes at all, and even those mentioned, such as sclerosis of Ammon's horn, would not explain the epileptic attacks.

Prognosis.—This should be made as to life and possible recovery or improvement. The life of an epileptic is not, as a rule, threatened by his attacks. Much more dangerous are the so-called accessories of epilepsy, the character changes, insanities and dementia. Much depends upon the severity or degree of the degeneracy, of the number of its stigmas; upon the frequency and severity of the attacks, the severity and nature of the exciting causes, as well as the age and the period of the commencement of the malady. The longer the disease has lasted, the more faulty the heredity, and the more numerous the attacks, the worse is the prognosis. According to Turner, cases beginning between 16 and 20 years show the greatest percentage of arrests of the attacks, while cases beginning before five are more dangerous, as they are prone to be confirmed, are the least amenable to treatment, and are liable to develop psychic defects (dementia). Remissions are very common, especially when the proper treatment is given. According to Habermass,[36] epilepsy is curable in 10.3 per cent. of patients. Freedom from intellectual defects was observed by him in 17.3 per cent. 21 per cent. were capable of earning a living; 30 per cent. were partially able to work, and 49 per cent. were not capable of doing any kind of work. The average duration of life is 25 years, and in 60 per cent. the epilepsy itself was responsible for death. The main danger is not so much the attack, as the accidents that may occur during it. Thus, being unconscious and helpless, the patient may drown, be burned, suffocated (by pillows during a night attack), or choked by vomited masses. He may sustain a skull fracture during a sudden fall on a hard pavement, or may die from an infected bitten tongue or lip. Therefore, the life of an epileptic is always subject to some danger. In this connection Spratling's statistics are very instructive. Out of every 100 deaths of epileptics, about four are the result of a single seizure, about 24 the result of status epilepticus, about 24 the result of some disease of the lungs, chiefly tuberculosis, about 12 the result of some accident, including suffocation in bed, about 10 the result of organic disease of the heart, and about 26 from other causes.

Even when the attacks cease, the prognosis is not always to be considered favorable, because the mental deterioration usually progresses in spite of the absence of the attacks. Psychologists, such as L. Pierce Clark, say that whenever the character is thoroughly and firmly fixed and is incapable of adaptation to work and social demands commensurate with the life level suitable to the individual's needs and capacity, a poor prognosis must follow. The maximum attention must be paid to eliminating or modifying the character faults which will permit the individual epileptic to objectivate fully his supernormal egotism, so that he at least may delay mental deterioration. The latter can be gradually followed up and determined by various psychologic (psychometric) tests, as outlined by Clark and Mileau.[37]

Course.—Epilepsy is essentially a chronic malady. It may exist for many years without incapacitating the patient to any great extent, though if untreated and in case the attacks cannot be brought under control, dangerous mental abnormal states may result, such as dementia, epileptic psychoses and idiocy. In the early years, the epileptic attacks may be frequent and disappear for a time, though the frequency of the attacks is not always responsible for the degree of mental deterioration. As a matter of fact, more or less pronounced signs of the latter, as well as character anomalies, may for a time precede the attack. Thus, the child is unusually irritable, quarrelsome, cannot concentrate the attention, suffers from insomnia, manifests groundless fears or anger, or it shows some sleep disturbances, such as night terrors, wets the bed, cries continually, is uncontrollable. Such episodes, however, do not always mean epilepsy. Then again, prolonged free intervals, remissions and states of apparent complete health are very frequent in this chronic disease, expecially when properly treated.

Equally unsettled is the question of the so-called pathogenesis of epilepsy, the mode of the occurrence and origin of the attacks or other phenomena observed during an epileptic seizure. Jackson's[27] and Schroeder van der Kolk's theory that the convulsions result from excessive discharges of nerve cells which "by morbid nutrition become of very high tension, of most unstable equilibrium," is replaced by the so-called cortical theory. According to this theory, the seat of the epilepsy is the entire cortex of the brain and possibly the cerebellum. It would probably be better to assert that the entire gray matter of the central nervous system is the seat of epilepsy, including the pons, medulla and spinal cord. The motor, sensory, and psychical symptoms occurring during the premonitory stage, the aura, and the post-epileptic states, all could very well be explained by the disturbed state of the cortex, and nothing else; the cyanosis and respiratory phenomena, by the involvement of the medulla or the bulbar centers, while the loss of reflexes can be explained only by the involvement of the spinal cord centers. The bladder incontinence is explained by the contraction of the bladder musculature, which is probably under the control of the sympathetic nervous system. On the other hand, experimental work on epilepsy shows that convulsions also occur in animals deprived of a cerebral cortex. Picrotoxin, absinthe, cinchonidin and other toxic substances may produce epileptic convulsions in such decerebrated animals, but they show only the tonic stage, the clonic phase being absent. In the decerebrated animals the medullary symptoms, such as the vascular and respiratory, precede the convulsions, and even the aura according to Pollock and Treadway,[28] thus indicating that the subcortical, not the cortical region is the starting point for the convulsions. As the pons and medulla are the only parts of the brain whose integrity has been found to be essential in the production of experimental convulsions, these portions, Pollock[29] says, must be considered the centers on the sites of discharge of epilepsy. Thus the old theory of Nothnagel[30] that the pons and medulla are the centers of epileptic attacks is again revived, in spite of the fact that this theory cannot, like the cortical one, so satisfactorily explain all, or even the majority, of the phenomena of human epilepsy, including the vasomotor disorders. The latter, according to Bechterew,[31] result from disturbances of the cerebral circulation which ultimately may cause epilepsy while according to L. J. Pollock both the circulatory and the respiratory changes occur as the result of medullary stimulation. Assuming irritation or some other lesion of the cortical centers as the basis of epilepsy, it is possible to offer an approximate explanation of every epileptic phenomenon in a human being. Asphyxia and cyanosis, for instance, can be ascribed to the immobilization of the chest during forced expiration at the moment of the tonic phase, in which all the muscles, including the abdominal group, participate. The blood is unable to return to the heart, and accumulates in the vessels, causing cyanosis. Pupillary dilatation and areflexia (loss of reflexes) are ascribed by Féré also to cortical lesion. Salivation is a mechanical escape of accumulated saliva in the mouth. The blood is the result of tongue biting or injuries to the mucous membrane of the mouth. The post-epileptic paralysis is ascribed by some, as Todd, A. Robertson, Hughlings Jackson, to the exhaustion of the cortical centers; by some, as Gowers and Löwenfeld, to their "inhibition;" by some, as Kussmaul, Tenner, and Féré, to their anemic condition. The latter theory is wholly untenable, as the brain during an epileptic fit is hyperemic rather than anemic, according to Bechterew[31] and Todorsky. Neither is the exhaustion theory satisfactory, as an exhausted cortex should cause not only an exhaustion of the motor centers, producing a paralysis, but also impairment of other functions, as sensibility, hearing and vision, which as a rule does not occur.

Attempts were made by Haig[32] to explain the epileptic attacks by poisoning the organism with uric acid and, by Krainsky,[33] with carbamic acid, and Haig even declared the possibility of controlling the epileptic attacks by instituting a proper metabolism. The excess of uric acid in the blood, he thinks, produces circulatory changes, affecting, in one direction, the intracranial circulation and producing the fit, and in another the stomach and intestines, bringing digestion and absorption to a standstill. Krainsky, on the basis of his experiences, concludes that the epileptic attacks are caused, not by retention in the blood of excessive amounts of uric acid, but by the periodic formation of ammonium carbamate. During the attack the latter breaks up into urea and water. In severe attacks and status epilepticus, the blood of an epileptic contains very large quantities of ammonium carbamate, while in the intervals its amount in the blood is very small. Unfortunately, such conclusions could not be confirmed (Hubert, Schnitzer[34]), though some authors, as Redlich, admit that an epileptic fit may be a reaction of the brain against some endogenous or exogenous poison. In conclusion, I should like to mention the psychologic theory of epilepsy as advocated by L. Pierce Clark, of New York. According to Clark,[35] a grand mal attack is the result of maladaptation of the epileptic to the surroundings. The patient reacts against them by a fit. The attack is thus an epileptic reaction, and has an individual, quite unique, psychology. When the defect of adaptation of the instinctive demands is hidden or repressed, the unconscious cravings are unable to gain satisfaction from existing conditions, and cause the psychobiologic break or loss of consciousness. Pure physical and chemical factors, he believes, cannot cause an essential epilepsy. For the latter, a defective make-up is necessary, which can be detected in an epileptic from early childhood, and upon this defective make-up is placed a variety of precipitating causes, which are varied as life itself.

From the brief review of the numerous theories of epilepsy, its pathogenesis, it can be seen how unsatisfactory this problem is at present, and it will remain so until histopathologic or biochemic researches help in settling this highly difficult and most important question.

Before proceeding with the discussion of the treatment of epilepsy, it would be proper to consider the other large groups of epileptic manifestations, known as epileptiform states or conditions. Here belong (*a*) Bravais-Jacksonian fits; (*b*) epilepsia continua (Koschevnikow's disease); (*c*) myoclonus epilepsy and (*d*) tuberous sclerosis (Bourneville's disease).

(*a*) **Jacksonian** (Bravais) epilepsy is the best known and most common type of *epileptiform* convulsions. The latter are morbid manifestations, which differ from typical epilepsy in the absence of the rapid and early loss of consciousness, of post-epileptic states, and, which is very important, in the presence of a definite etiology. The convulsions in epileptiform fits are usually unilateral, gradually becoming generalized and universal, when loss of consciousness also obtains. The marked differences between epileptic and epileptiform states can be distinguished only in typical cases, while less typical forms may show features peculiar to either. For instance, genuine epilepsy may be strictly unilateral, while loss of consciousness may occur also in Jacksonian epilepsy. Tumors of the brain, usually causing the epileptiform type, may in some cases give a typical picture of genuine epilepsy. In children, this is especially true of so-called organic hemiplegic epilepsies, that is, epilepsies associated with cerebral infantile hemiplegia. Clinically, they show a typical genuine epilepsy, pathologically, an equally typical organic brain lesion. And it frequently happens that an epileptic child, upon careful study of the history, and course, is ultimately found to be not an epileptic at all, but a sufferer from some brain lesion complicated by epileptiform states. Wherever situated in the brain (frontal, occipital, parietal, or in other areas) a focal lesion may bring on an epileptic attack. Such patients usually show focal symptoms first, later followed by general convulsions. Though this helps the diagnosis of organic epilepsy, the focal symptoms are often mild and little in evidence, while the general symptoms are severe and marked. In such cases, the symptoms of generalized epilepsy and even of status epilepticus may prevail. Anatomic causes of such etiologically obscure epilepsies are the same as in infantile cerebral hemiplegia, such as porencephaly, developmental anomalies of the cerebral convolutions, lobar sclerosis, foci of encephalitis, hemorrhage, softening and embolism.

Epileptiform states are also frequently caused by syphilitic brain lesions, general paresis, multiple sclerosis, tumors, abscesses, hydrocephalus, and meningo-encephalitis. The epileptiform manifestations of these conditions form only one group of symptoms. Frequently, these manifestations are so marked that they dominate the clinical picture in the form of Jacksonian convulsions.

Symptomatology.—The cardinal features of Jacksonian epilepsy are localized twitchings or clonic convulsions confined to one limb, to a portion of the latter, or to the face. The twitchings either remain stationary or spread, according to the course of the cortical centers which have a definite localization. The convulsions, starting, for instance, in the center of the foot, spread to that of the leg, thigh, arm and face; or starting in the face, they successively invade the arm and leg, or when starting in the arm, they may involve either the face or the leg, and may spread over the opposite side where they will involve the leg, arm and face. They may also start in the toe, finger or shoulder, always spreading according to the localization of the functional cortical areas, successively affecting, but never dodging any of them. For instance, convulsions starting in the leg never jump over or miss the arm, before involving the face, or from the face they never jump to the leg. Usually no aura precedes Jacksonian convulsions, consciousness, as a rule, being retained; when it is lost, the twitchings become generalized. The patient witnesses his fits which he often can stop artificially, by ligation of the arm, for instance, if the affection started there. Equally unusual are the initial cry, falling to the ground, cyanosis, foaming and post-epileptic sleep. The muscular contractions are mainly clonic—not tono-clonic as in genuine epilepsy, but in rapid jerkings of a few seconds or minutes' duration, often followed by a temporary paralysis of the affected limb, lost or exaggerated reflexes and a positive Babinski reflex. In the same patient the motor restlessness always starts in the same organ, an important factor, indicating with precision the exact seat of the brain lesion.

The nature of the lesion can sometimes be determined by a careful study of the patient's history, by serologic and ophthalmoscopic studies. Syphilis or subdural hemorrhages, can be diagnosed with great precision.

Like epilepsy proper, Jacksonian epilepsy shows atypical types. Instead of purely motor there may be sensory manifestations, which will follow the same cycle as the motor type. For instance, the patient may complain of paresthesias (numbness and pricking) or pain in one arm, one leg, half of the face, tongue, body, or even in one finger, or toe, and these sensory manifestations may spread following the same route as pure motor symptoms, that is, from the toe to the leg, arm, face, or *vice versa*. The result may be a progressively developing hemi-anesthesia, which may be complicated, afterward, by progressive involvement of the motor centers (acute progressive hemiplegia), as well as by aphasia, hemianopsia, agraphia, alexia.

The **course** and **prognosis** of Jacksonian fits depend upon the etiologic factors. When their removal by medicinal or surgical treatment is possible, then complete recovery or improvement of some of the distressing symptoms (headache, threatening blindness, etc.) may be expected.

(b) **Epilepsia Continua** (Koschevnikow[38]).—In this form of epilepsy the epileptiform convulsions are uninterrupted; do not stop even during sleep, and last for hours, days or months, unless stopped by death or treatment. An entire half of the body, one limb, or portion of a limb is involved in involuntary clonic contractions, jerkings, twitchings, without an initial cry, fall, tongue biting, or post-epileptic states. Such cases could be designated as Jacksonian status epilepticus, analogous to the classic status epilepticus. In the latter, however, elevation of temperature, and unconsciousness, are present, and what is still more characteristic, intervals occur free from convulsions. Probably some cases of continuous epilepsy have been described under the name of Jacksonian status epilepticus, for instance, by Bernheim, who observed 95 convulsive attacks in two hours in one patient. Jacksonian status epilepticus cannot be called continuous epilepsy, in which there is but one attack lasting for a very long time without inconveniencing the patient. The general condition may suffer very much, depending upon the underlying cause, which may be syphilis, abscess, tumor, encephalitis, or some other organic cerebral lesion. Cases of continuous epilepsy are by no means frequent, only a few having been reported in the literature, chiefly by Russian neurologists (Koschevnikow,[38] Orlowsky,[39] Choroschko[40])—all in adults.

(c) **Myoclonus Epilepsy** (Clark and Prout).—Continuous epilepsy somewhat resembles so-called myoclonus epilepsy—a name given by Clark and Prout[41] to a combination of myoclonus and epilepsy.

The former, described in 1881 by Friedreich as paramyoclonus multiplex, consists of rapid, lightning-like contractions of one muscle, a group of muscles or a portion of a muscle. In the latter, instead of marked coarse muscular contractions, there are mild fascicular or fibrillary twitchings affecting both the protagonists and antagonists. When the large muscles are affected, especially in advanced cases, the myoclonic contractions may be followed by a locomotor effect, such as flexion, extension, pronation and rotation of the leg or arm. The extremities and trunk are usually involved, while the larynx, facial muscles, respiratory and ocular muscles are spared, or may be affected only in advanced stages. The contractions increase from psychic and physical exertion, such as turning in bed, and cannot be imitated or reproduced at will. The myoclonic contractions may be *combined* with epileptic attacks; may *precede* them for a long time or, as is usual, may *follow* them. In other words, myoclonia may either occur as an isolated disease or be associated with epilepsy, preceding or following it. According to Clark and Prout, in a combined myoclonia and epilepsy, the epileptic attacks are not typical. Aura, the tonic stage, tongue biting and post-epileptic coma are absent. The myoclonic epilepsy usually appears in childhood, in 75 per cent. of cases, according to Clark and Prout, occurring between the ages of 9 and 15 years; the entire life of the patient thus being made up of myoclonic and epileptiform attacks, of which the former predominate. There are, as Lundborg[42] points out, good and bad days, but psychic deterioration, feeblemindedness, is progressing and ultimately, with very few exceptions, results in dementia. Lundborg, therefore, prefers to call it progressive myoclonus epilepsy, and points out its possible occurrence as a family disease.

Diagnosis.—This is not difficult, in view of the characteristic combination. Its difference from continuous epilepsy is also evident.

The **prognosis** is exceptionally bad, myoclonic epilepsy being the worst type of epilepsy, for insanities and idiocy are the usual accompaniments and the disease itself may last until an old age.

Treatment.—Hospital or institutional care is absolutely indicated.

(d) Tuberous Sclerosis Epilepsy (Bourneville's Disease).—Another sad complication of epilepsy is tuberous sclerosis, in which epilepsy is combined with idiocy. Described chiefly by Bourneville,[43] alone and also in connection with D'Olier[44] (in 1880), this form of epilepsy is of particular interest, especially to pathologists, in view of the great wealth of brain changes to be found in such epileptics. It may be said that this is the only type of epilepsy with a definite pathology. It is a rare clinical entity: Only 10 cases were found by Bourneville in 25 years among his enormous clinical material. The clinical picture is made up of epilepsy, idiocy, and skin lesions in the form of small wart-like tumors covering the face and cheeks, especially around the nose. It is noteworthy also that they are present in the kidneys and heart. These tumors are nevi sebacei, fibro-myomas and angio-fibro-lipomas. The disease begins in early life when either epileptic fits or psychic defects first appear, gradually increases and ends in dementia or idiocy. In some cases the intellectual decay is rather mild, but instead moral defects (moral insanity) are pronounced (inclination to steal, lying). The epileptic attacks are usually generalized, occurring as ordinary epileptic fits, but they may also be local and followed by paresis, paralysis and contractures. The clinical picture can be explained by the anatomic lesions in the brain, in which there are dense, hard, nodular masses, somewhat elevated and irregularly scattered over the brain tissue. The islands are distributed in various convolutions of the cerebral cortex, Ammon's horn and the large basal ganglia, while the centrum ovale, cerebellum, pons and medulla are spared. The nodules consist of glia tissue and a great number of large atypical ganglion cells; the upper layer of the cortex, the so-called stratum zonale, is greatly increased in size, and in other layers, the ganglion cells are usually destroyed, vascular and pial changes being absent.

The **diagnosis** of tuberous sclerosis can be made from the nodules in the skin, the history, and the early occurrence of mental and other psychic defects. Even in the absence of skin lesions, according to H. Vogt, a tuberous epilepsy may exist. The *idiocy* is a sufficient index of the severe structural brain lesions, which explain very well the clinical symptomatology—the epilepsy, and the psychic defects. It also explains the progressive course, the rapid onset of idiocy and the hopeless prognosis, as many portions of the brain substance are destroyed and replaced by islands of sclerotic tissue. The remarkable feature observed in cases of tuberous sclerosis is the possible occurrence of moral instead of intellectual defects. Both, of course, are due to the anatomopathologic changes in the brain, which in tuberous sclerosis are more evident than in any other type of epilepsy, except possibly some Jacksonian forms. It is not surprising, therefore, that on account of anatomic brain deficiency some epileptics frequently come into conflict with the law or environmental conditions, in the case of children, for instance, with the school authorities.

Epilepsy may undoubtedly be found among criminals, morons or other moral perverts. Whether every criminal should be held responsible for his deeds must depend sometimes upon his physical or mental condition; that is, upon whether or not there is hidden in him some mental disorder, such as epilepsy, which may influence the abnormal conduct. Epilepsy, therefore, is not only of great medical importance, but also of pedagogic and medicolegal value.

General Treatment of Epilepsy.—In every case of epilepsy the possibility should be considered of its being a symptom of some organic brain lesion, or of some lesion of organs more or less remote from the brain (reflex epilepsy,) or of some constitutional disorder (uremia, diabetes, uric acid diathesis). In such cases, the treatment must be confined to the etiologic factor, that is, it should be individualized. When the diagnosis of epilepsy is finally made, inquiries into the patient's heredity, family history, habits, mental and character make-up should be made, and the habits of life should, if necessary, be corrected. L. Pierce Clark justly insists that our main concern in the treatment of the the epileptic should be his primary make-up, as it is a defect which antedates the seizure phenomena by years. In other words, the primary character endowment of the epileptic must be considered. An epileptic should be placed in surroundings suitable to his character, and inclination. A dietetic regimen with regulation of sleep, meals, bowel function, physical work, exercises, sexual life, and all should be looked after in each case. A meatless diet is advocated by some, as Cramer,[45] while avoidance of physical and mental overwork and strain, plenty of sleep, with exclusion of psychic excitements, should always receive proper attention. In fact, the regulation of the patient's mental and physical condition is of more importance than the medicinal treatment, which is but a supplement to the foregoing measures, and should be resorted to only when the attacks or their equivalents are too frequent and severe. Of the medicines, bromids still occupy the first place. Sodium, potassium, ammonium, strontium salts, alone or combined, are of great benefit, depending on the dosage and the individual case. If the attacks usually occur during the night, a single dose of 5 to 10 gr. to a child from 4 to 10 years of age is sufficient. In diurnal attacks, a repeated dose of the same amount or, if necessary, larger doses, should be given immediately before meals. A five year old child may receive 2 to 4 gm. of bromids a day; a 10 year old, from 4 to 6 gm. Binswanger[46] recommends larger doses, from 3 to 6 gm. a day, to a five year old child.

The bromid treatment should be kept up for many months or years, the dose being gradually increased or decreased, according to the indications. Of these, the most common are skin eruptions (acne bromatum), digestive disorders, muscular weakness and drowsiness. Some authors, as Gilles de la Tourette, give bromids till the pupils begin to react slowly, others till the pharyngeal or corneal reflexes disappear. The dose may be reduced if table salt (sodium chlorid) is removed, or its amount reduced in the food. Richet and Toulouse advocate such a treatment with a salt-free diet, recommending as a food the use of bromids instead of chlorids. The dose of bromids, if sodium chlorid is excluded, may be reduced to 1 or 2 gm. a day. In the absence of sodium chlorid in the food, the bromids are much better absorbed by the tissues; but, when sodium chlorid is used, the latter, on account of its greater chemical affinity for the tissues, is preferably absorbed, pushing the bromids out, so to speak, as Laufer remarks. Bromids are also given in combination with opium (Flechsig's method), or with codein and Adonis vernalis (Bechterew's method). Neither of these methods is used in treating children, especially Flechsig's method, which is better adapted to sanatorium or hospital cases. In cases in which the bromids cause severe skin or other complications, they may be replaced by zinc preparations and luminal. Zinc may be administered in the form of zinc valerianate or zinc lactate, either alone or combined with extract or tincture of belladonna. Echeverria[47] asserts that if bromids are given with Fowler's solution, in one mixture, the skin accidents can be prevented; but Fowler's solution is also used alone for the treatment of the brom-acne. This usually subsides when the bromids are discontinued. For the time being they can be replaced by belladonna or zinc preparations. Luminal is usually used with adults in 1½ gr. doses, in tablets or capsules, given once, or, if necessary, twice or three times a day. In many cases this remarkable remedy proved far superior to bromids; in some it was without effect. In children, five years of age, the dose is about ⅔ or ¾ gr. once a day.

The Treatment of the Attack Itself.—In the stage of the premonitory symptoms and aura, the patient usually retires to some safe place (children place themselves on the floor) as if instinctively trying to protect themselves from possible injuries. In auras occurring in partial or organic epilepsies, the patient often succeeds in preventing the attack by tightly ligating the involved limb, the arm, for instance. During the attack itself, the patient should be made comfortable, the collar should be loosened, the tongue prevented from being bitten (by a cork or spoon being placed between the teeth), the head protected from bumping against the floor during the clonic stage. Any active interference, such as morphine injections, chloroform or hot baths, are unnecessary, though in attacks of eclampsia recourse is frequently had to the latter two remedies. They certainly do no harm. In the post-epileptic stage, the patient should not be aroused or disturbed, as suppression of post-epileptic sleep, according to Féré, may cause disagreeable after-effects (persistent and painful headaches).

In serial attacks and status epilepticus, the same rules should be observed, though, in view of threatening exhaustion, inhalations of chloroform, chloral (by rectum), morphine (hypodermically) must sometimes be given.

In obstinate cases, in which the attacks are unusually frequent or uncontrollable, or in which the mental symptoms are marked, hospitalization is strongly indicated. Epileptic colonies, which are located in some states, at Sonyea in New York (Craig Colony) and at Dixon, Illinois, are the most rational institutions. In these the epileptics are educated, guided, taught manual training, gardening, dairying, farming, casket making, and carving. (Spratling).

As many epileptic children do not show marked mental defects, attendance at school should not be forbidden, unless the attacks occur during the day, and unless they can be definitely proved to be aggravated by mental effort.

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HYSTERIA AND SOMNAMBULISM

Hysteria is a typical functional nerve disorder, or rather a peculiar complex of various nervous and psychic disorders. By some authorities, Bernheim,[1] for instance, it is not an autonomous disease at all, but rather an epiphenomenon, an appendix, as it were, to diverse maladies. It may be said that hysteria is an abnormal mental state, a psychoneurosis, characterized by excessive suggestibility and highly vivid reactions on the part of the psychic sphere to slightest stimulations, purely traumatic or psychic.

Etiology.—The causes of hysteria are, like those of epilepsy, predisposing (remote causes) and exciting (immediate causes). To the former belong defective heredity (mostly direct) and unsound social and domestic surroundings. Exciting causes are of the nature of socalled shocks, sudden emotions or psychic traumas, chronic constitutional diseases, such as tuberculosis, syphilis or intoxications (alcohol, lead poisoning). A combination of both predisposing and exciting causes is instrumental in the causation of hysteria. All are equally subject to this disease, whatever the social status, race, age, sex, or the climate in which they live. Females are more frequently affected than males. The youngest patients whose cases are recorded are those mentioned by Scheffield[2] (1½ years old), Bruns (three years), and Cramer[3] (four years).

Symptoms.—The numerous symptoms of hysteria are usually classified as permanent or authentic and accidental or additional.

The permanent symptoms are generally considered specific for this disease and, therefore, are called stigmas. Accidental symptoms may simulate any organic nerve or visceral lesion, and may be present or absent. The symptoms of either group vary in any case according to their number, type and intensity, being as a rule mobile, that is, changing—coming and going. In children, hysterical symptoms are often represented by one symptom only—monosymptomatic hysteria. Furthermore, the symptoms of hysteria in children are not always typical. They are rather rudimentary, not fully developed, although otherwise they possess the same features as in adults. Therefore, they will be given only a general review here, and the special features alone, occurring in children, will be emphasized.

The *stigmas* of hysteria are sensory, motor and psychic.

Sensory stigmas appear in the form of (*a*) anesthesia (complete loss of all sensations), (*b*) hypesthesia (incomplete loss of sensibility), (*c*) partial or dissociated anesthesia, (*d*) increased sensibility-hyperesthesia, and, which is rare, (*e*) abnormal sensibility-paresthesia. Usually, in children, all these five types of sensory disturbance are absent, or at least are much more rarely observed than in adults.

The most remarkable type of sensory disturbance is *hemi-anesthesia*, that is, anesthesia confined to an entire half of the body, including the face, trunk and the mucous membranes of the eyelids, cornea, nose and mouth.

In addition, the higher sensory organs, on the same side, like those of smell, taste and hearing are simultaneously affected. Vision on the affected side shows a contracted field either for all colors, in their normal order (white, blue, yellow, red, green and violet) or for only certain colors, resulting in overlapping of the color fields. For instance, the field for blue is normally larger than that for red, but it may in hysteria become so much contracted that it will appear smaller than that of the red; the latter thus will overlap the former. In severe cases the contraction may be so great that no color perception is apparent (achromatopsia), objects appearing gray or discolored.

In less severe cases, some complementary colors are still perceived. For instance, when a person looks at yellow he will perceive red, and *vice versa* (dyschromatopsia). The same anomalies may obtain in the senses of smell and taste, odors being either totally unperceived, the strongest substances lacking exciting power, or their action may be reversed, bitter tasting sweet, and foul odors being agreeable. Hearing may be totally abolished, diminished or excessively acute. Charcot[4] says that in case the hemi-anesthesia is bilateral, the foregoing sensory phenomena will also be bilateral. Another feature of hysterical hemi-anesthesia is that it is strictly confined to one-half of the body, usually the left, being divided from the normal half by a straight line. Thus, half of the forehead, nose, lips and trunk will be insensible to pin pricks, touch, temperature, pressure, muscle and position senses; the other half being normal. The dividing line is frequently the mesial plane of the body, but occasionally it may be displaced ½ inch to either side of the mid-line. It is always a straight line.

The anesthesia often involves but one limb or a segment of it. In other words, it may occur as monoplegia or segmental anesthesia. In any case, the region of anesthesia ends abruptly. Segmental anethesias involve only the hands, forearms, feet or legs, producing what is known as glove and stocking anesthesia. Charcot calls this anesthesia in the form of geometrical figures. It may affect both lower or both upper arms, causing a paraplegic type of hysterical anesthesia, with all its peculiar characteristics—the preservation of motility, absence of atrophies, joint and skin lesions. Finally, it may be irregular, in the form of patches, spots and islands, scattered over various portions of the body. Whether unilateral or segmental the anesthesias are frequently incomplete, or dissociated. For instance, temperature sense may be less involved than that of pain; or the touch may be more involved than the muscle sense. Strongest currents, galvanic or faradic, are borne without pain, indicating also loss of electrocutaneous sensibility. Janet[5] holds that in spite of such profound sensory disturbances, the patient does not seem to experience any inconvenience. The totally insensible arm is not burned or otherwise injured, as commonly occurs in organic anesthesias, caused, for instance, by syringomyelia; and, in spite of the enormously contracted visual field, the patient has no difficulty in finding his way in the most crowded places. In fact he does not even know that he has any sensory disturbances until his attention is called to them, and only in very rare cases does he experience discomfort, numbness, for example, in the anesthetic limbs. In other cases, as Lasègu pointed out, some motor disorders are present. The patient, for instance, is able to move the anesthetic limb only when he watches it, and he must look at his hand when he attempts to move it. Whatever the intensity, type or localization, hysterical anesthesia differs from an organic form, caused by peripheral nerve, root, spinal cord or brain lesion, because it does not follow the distribution of the nerves or the roots, and is not associated with reflex anomalies, bladder and other disturbances (trophic, vasomotor), which are considered peculiar to organic lesions of the nervous system. Other peculiarities of hysterical anesthesia are their disappearance during sleep, from which a patient can be aroused by a painful irritation, and the possibility manifest to transfer them from one part of the body to another (transfer phenomenon). The totally insensible skin is not changed in color, and does not bleed when pricked by a sharp pin. That the foregoing disturbances of sensibility are exceedingly typical of hysteria is generally admitted. Their pathogenesis is unsettled; some authors, as Bernheim,[6] assert that these sensory phenomena are artificially produced, either by autosuggestion or, as Babinski believes, by a suggestion on the part of the physician. Whether produced artificially, by the patient himself or another, the presence of the sensibility and sensory disturbances, as noted, speaks for hysteria alone. This is also true of hypesthesias—incomplete loss of sensibility. In some cases of hysteria the opposite phenomenon is present, namely excessive pain (hyperesthesia), confined to small areas of the skin, most commonly in the region of the ovaries ("ovarie"), the center of the inguinal region in men, spots under the breasts, over the forehead, sternum and many other areas. Either a slight touch, or rubbing of these areas, or a more or less strong pressure may cause intense pain and even provoke a general convulsive attack. Therefore they are called hysterogenetic zones. Bernheim considers them as artefacts, the result of suggestion, and says that they can be produced at will. Paresthesias, such as pin-and-needle sensations, burning and numbness are rare in hysteria. Pain however, is common, being either spontaneous, a neuralgia (trifacial sciatica), arthralgia (pains in and around the joints), visceral neuralgia (pain in the stomach and intestines), or headaches (hysterical clavus). The latter are usually circumscribed, confined to one spot, occurring from physical emotions or from excessive attention given them.

Motor disturbances are usually associated with sensory changes. In mild cases they occur as tremors and myoclonic contractions; in severe cases, as paralysis, contractions and convulsions. Tremor may affect any muscle in the body, being regular, rhythmic, from 1 to 13, usually from five to six, contractions per second. It increases from psychic emotions, persists during rest or may be continuous, and ceases during sleep. It may occur in attacks of half an hour's duration, usually brought on by some excitement, and affecting portions of the body covered with anesthesia. Myoclonia is a severer type of tremor, the excursions of the contractions being greater, their amplitude wider, often resembling such acts as hammering, dancing, jumping and saluting (hysterical chorea). *Hysterical paralyses,* in their localization, resemble the hysterical anesthesias, being as a rule combined with the latter. Thus, the entire half of the body may be paralyzed (hysterical hemiplegia), or only one limb (hysterical monoplegia), or two limbs (upper or lower)—hysterical paraplegia. In any of these types, the paralysis is always confined to an entire segment, that is, a *portion* of an extremity. It may involve the *entire* limb, but never occurs as an isolated muscle paralysis. In other words, a paralysis of a single muscle does not occur in hysteria; such paralysis always indicates an organic nerve lesion. But organic lesions of the nervous system also cause hemiplegias, monoplegias and paraplegias and these may be simulated by hysteria and should be differentiated from it. In hysterical hemiplegia the paralyzed side is anesthetic, which is rather unusual in an organic hemiplegia. If the latter is combined with a hemianesthesia, hemi-anopsia occurs, tremor in the form of hemi-chorea, hemi-athetosis and other signs typical of the so-called thalamic syndrome.

Excluding the anesthesia, an hysterical hemiplegia is never associated with paralysis of the seventh or other cranial nerves (third, sixth, twelfth). The muscles supplied by these nerves may show abnormal deviations which, however, are due to their spasmodic states, that is, to muscular spasm or contractures, but not to paralysis.

In hysterical hemiplegia the reflexes are either normal, exaggerated or diminished, with a normal plantar response (no Babinski, Oppenheim, Chaddock or Gordon signs). In organic hemiplegia, the reflexes on the paralyzed side are exaggerated, as a rule, with positive Babinski, Oppenheim, Chaddock and Gordon signs.

In hysterical hemiplegias, foot and ankle clonus are almost always absent, (though Charcot reported such cases with typical clonus). In organic hemiplegia, clonus is frequent.

Many other signs differentiate organic hemiplegia from hysterical. For instance, the fingers' sign of Souques occurs only in organic hemiplegia. Elevation of the forearm (passive or active) is accompanied by extension and abduction of the fingers because of the contraction of the interossei muscles. The Klippel-Weil sign occurs in organic hemiplegias with contractures: passive extension of the fingers or toes is followed by flexion of the patient's thumb or great toe. In hysterical lesions, the same act causes extension of the thumb and great toe. In hemiplegias without contractures, Raimiste's sign is of great value. In this the patient's hand and forearm are slowly elevated to a vertical position. On removal of support the hand droops, forming an angle of 130 or 140°. This never occurs in healthy or hysterical patients, but appears early in organic lesions, and during coma it is thus possible to ascertain the exact site of the lesion. Another of Raimiste's signs is that associated movements of abduction and adduction obtain in the paralyzed limb, when the healthy one is abducted or adducted under resistance. This does not occur in hysteria. In testing for Néri's sign, extension of the leg, when the thigh is flexed over the abdomen (Lasègue sign), will be possible only to 40 or 50°, while, on the healthy side, the same procedure will bring the extended leg to an angle of 70 or 75°. Of many other signs described for differentiation of hysterical from organic hemiplegias, I will but mention Hoover's complemental opposition sign.[7] The physician places his hand under the heel of the healthy foot and orders the patient to raise the paralyzed leg. Lifting it will cause a strong pressure by the healthy heel on the physician's hand. When the hand is placed under the paralyzed heel, lifting of the healthy leg will cause no pressure on the underlying hand. In hysterical hemiplegias, as first pointed out by L'hermitte,[8] the opposite obtains, that is, attempts to raise the paralyzed leg do not cause pressure exerted by the opposite, healthy heel, provided the hysterical hemiplegia is not flaccid. Of all these signs, Babinski's is most valuable. It is an easy test, and does not require the patient's coöperation, which is necessary in some other procedures.

Hysterical hemiplegia can be differentiated by many other features. For instance, the *gait* in hysterical hemiplegia is totally different from that in organic hemiplegia, in which the paralyzed limb is circumducted, the foot drooping somewhat. In hysteria, the patient drags the paralyzed leg as if it were a dead, useless appendage. He scarcely bends the hip, knee or ankle joints. Paralysis of a side in organic hemiplegia is sooner or later followed by atrophy (from disuse) or joint lesions. These are absent in hysteria.

In hysterical *monoplegia* anesthesia is simultaneously present. It involves the entire limb, ends abruptly and is limited by a circular line perpendicular to the long axis of the body or limb. A combination of paralysis, with anesthesia of one limb, also occurs in cortical brain lesions. In the latter, however, the anesthesia does not end abruptly, but gradually diminishes from the periphery to the center. The reflexes and tonicity of the muscles also are exaggerated in the organic variety, remaining normal or diminished in hysteria.

In hysterical *paraplegia* differential diagnosis is the same as in hemiplegia. Clonus, Babinski's phenomenon, and other signs of a spinal cord lesion (trophic, vasomotor, bladder and other disturbances) will be absent. Contractures may exist in the foregoing types of hysterical paralyses, that is, a limb may be not only paralyzed, or anesthetic, but also contracted. The hysterical contractures may follow either some physical or psychic trauma, or electrical stimulation will make them more pronounced. They involve single muscles, as those of the eyelids (blepharospasm), or a group of muscles, such as the flexors or extensors of an extremity. During sleep the contractures usually disappear, though often they persist even during the most profound sleep. They may be relaxed, though not always, during an artificial narcosis. If deep narcosis fails to cause relaxation, one should suspect adhesions as the cause of the immobility of the affected joints. If there are no adhesions, atrophies, electrical changes and reflex anomalies are absent in hysterical contractures.

The most striking motor disorder in hysteria is convulsions. As in epilepsy, they occur as grand or minor attacks, the latter being much more frequent than the major spasms, which, in their classical form, occur exceedingly rarely. As described by older authors, especially Charcot, they consist of several stages: (1) prodromes, usually merging with an aura; (2) epileptoid stage; (3) stage of contortions and excessive movements, and (4) stage of delirium.

The prodromes and aura may be of the same character as in epilepsy. The most usual aura is the so-called hysterical globus, a choking sensation as of a ball rolling up to the larynx from the stomach. Instead of globus, there may be sensory visual manifestations (hallucinations, flashes before the eyes), auditory (whistling, buzzing, various noises), purely psychic phenomena (causeless laughing, crying, sadness or gaiety). All these morbid manifestations are usually preceded by some emotional disturbance (shock). Frequently the attacks are confined to these premonitory symptoms, being an attenuated or an abortive form of hysterical seizures. In rarer cases, the aura is followed by a fall, not sudden, as in epilepsy, but, rather, deliberate, a gradual sinking to the floor, the patient not being injured as is often the case in epilepsy. There may be a cry or rather a series of cries, followed by the epileptoid state. In the latter, tonic and clonic contractures affect either one extremity or the whole body, without cyanosis, while sensibility, reflexes, pupillary reaction and consciousness are usually preserved. The clonic and tonic contractures lead to abnormal postures, such as arching of the body forward, backward, laterally (opisthotonos, emprosthotonos, lateral opisthotonos), followed in a few minutes by excessive movements or contortions (stage of clownism), which, according to Bernheim, are very frequent in children. In this stage, the patient performs wild, excessive movements manifesting joy or terror; he rolls around, stands on his head, assumes threatening attitudes, and finally enters the last stage—that of delirium. Often he falls asleep, or the attack recommences. The entire attack may endure for hours, but usually lasts from 15 to 30 minutes. Its duration is much longer than in epilepsy. These attacks, like those in epilepsy, occur in series, and, in analogy to status epilepticus, a so-called *status hystericus* has been described. Thus Marie and Souza-Leite[9] observed in a 31 year old hysterical patient as many as 1636 successive attacks. The first series comprised altogether 4506 attacks, the minimum duration of an attack being 12 minutes, the maximum, 22 minutes. The second series comprised 17,083 attacks, the duration being from 16 to 50 minutes. The minimum daily number was 176. LeGrand du Saull[10] recorded 21,708 attacks in 25 days. As in epilepsy, the attack does not pass through all stages. It may, as previously noted, be confined to the aura, or to the epileptoid stage; it may be represented by the stage of clownism, or by the final stage—attacks of sleepiness (narcolepsy or lethargic states). These will be discussed elsewhere. In other words, in these seizures, a classical major hysterical attack may be reduced to its elements. For instance, there may be violent shaking of the whole body or of a limb, or there may be merely outbursts of anger, excessive laughing or crying, or delirium with hallucinations and illusions, or manifestations of clownism, the assumption of passionate attitudes with violent tossing or struggling, or tonic or clonic spasms without falling, crying, etc. Whatever the type, violence, or duration of the attack, the patient is not exhausted as in epilepsy. As Janet says, he feels rather comfortable and remembers little or nothing of what occurred during the seizure. In other words, the patient has an amnesia. Janet even goes so far as to warn against a diagnosis of hysteria in cases in which violent agitation occurs with no loss of consciousness, and with an accurate remembrance. Such cases, according to Janet, are not hysteria, but are of the nature of so-called *psychogenetic convulsions.*

Typical as the convulsions may appear in hysteria, epilepsy and so-called psychogenetic states, a differential diagnosis is not always easily made, especially in children, in whom the convulsions, like other hysterical phenomena, are usually rudimentary. The history is, of course, of great value. The younger the child, the less likelihood is there of hysteria, according to Abt's[11] observations, and the greater likelihood of epilepsy. An hysterical child is, in general, active, wide-awake, intelligent, bright, a good pupil at school; but an epileptic child is less intelligent and rather apathetic. The epileptic attacks are alike, while the hysterical attacks are mobile and changeable (Bézy).[12] The epileptic attacks are independent of will or suggestion, occurring irrespective of place and surroundings, accompanied by total loss of consciousness and involuntary motions; hysterical attacks are usually of emotional origin or the result of suggestion, the consciousness often being retained to greater or less extent. Other features peculiar to either type are outlined in the article on epilepsy, to which the reader is referred.

While many authors look upon the attacks as the result of some emotional disturbance (Raimiste, for instance), others, as Hartenberg,[13] consider them as a manifestation of weakness, helplessness, caused by disagreeable recollections or autosuggestion, in short, as a reaction of defense. A third group of authors, especially Janet, consider the hysterical attacks as an aborted state of somnambulism. To the latter are also referred *dreamy* states, or dreams, and abnormal sleepiness, the characteristic feature of all these states being *amnesia*, oblivion of everything that occurs during them. Another feature is that the attacks, or the patient's actions, are repeated with remarkable regularity and stereotypy in somnambulism. In short, somnambulism, as defined by Janet,[14] is "an abnormal sleepy or dreamy state in which an individual acts and thinks without recollecting when the condition is over what was going on in his life during that condition. He reproduces in his dream the emotions he experienced, like biting by a dog, some injury, etc., and lives again through the events of his life, with its former emotions, actions and thinking." The unconscious ideas pop up during somnambulism, replacing the consciousness for a certain period of time.

During somnambulism, the patients display great activity, although they pay no attention to the surroundings. They do not answer questions and do not recognize any one's presence. Upon awaking, they resume their ordinary activities, with no recollection of the events that occurred during sleep. These conditions may last for long periods and may occur as ambulatory automatism or fugues with the same characteristic features as described in epilepsy. Janet believes that many fugues observed in epileptics are really hysterical manifestations.

Another consequence of somnambulism is what is known as double or dissociated personality or double existence. There is a normal existence, and another—abnormal—condition. The events of the former have nothing to do with those of the latter. For instance, in the normal state a person may be depressed, unable to fix the attention or do any kind of physical or mental work; in the state of somnambulism, the patient's condition may totally change. Excitation may replace depression, the mental and psychic faculties undergoing remarkable transformations. Janet records a case of a patient who forgot in one state how to read and write, the lost faculties returning with pristine vigor in the other state. The mental functions are never wholly lost in either state, though they are performed somewhat absurdly, and without the will or coöperation of the patient. The abnormal states can also be reproduced artificially, by hypnotism, which is considered to be a state of somnambulism.

Somnambulism must be differentiated from other similar states, such as narcolepsy and lethargy. In narcolepsy the attacks of sleep are short, in the latter, lethargy, they are prolonged, sometimes extending over long periods. Both are characterized by an irresistible desire to sleep, irrespective of time and surroundings. The patient may fall asleep on the street, in school, during occupation or play. The awakening is accompanied by fatigue, general malaise, sometimes headache and amnesia. The pupils are contracted during sleep, the extremities relaxed, the lids closed and tremulous; respiration is accelerated; the face slightly flushed; the general sensibility abolished. The attacks of sleepiness may be preceded by an aura—ringing in the ears, heaviness in the head, dizziness—and may follow or replace an hysterical attack of convulsions (Bernheim, Parmentier).

The symptoms of hysteria—anesthesias, paralyses, contractures, contracted field of vision, convulsions, somnambulistic states—are generally looked upon as stigmas or permanent signs. Janet, however, considers only the somnambulism as an hysterical stigma, while others, as Bernheim and Babinski, consider these signs as artificial, the result of suggestion or autosuggestion. However, if present, these symptoms are highly characteristic of hysteria, and nothing else.

The stigmas are usually associated with a great variety of so-called accidental symptoms. To these belong disorders of gait, speech, vision, gastro-intestinal and genito-urinary tracts, skin, vasomotor system, and so forth. Some of the accidental symptoms are quite typical and warrant a brief description.

The disorders of gait in hysteria take usually the form of astasia or abasia. In astasia the patient is unable to stand; in abasia, he is unable to walk, in spite of the fact that the voluntary movements, the power, the sensibility, and the reflexes are undisturbed. While in a recumbent posture the most complex movements are possible, but they are impossible when standing or walking, though the patient may be able to climb or run. Another type of hysterical gait is spasticity in walking, as in an organic paraplegia, in which the patient takes small steps scratching the feet along the floor, the legs being but slightly flexed, and carried along as inert bodies. In noting these types—astasia, abasia, and spasticity—the impression is given that the patient has forgotten the art of walking. His efforts much resemble those of an infant who while lying can move the legs in various directions, but cannot stand or walk. In the second (spastic) type, the gait is clumsy, imperfect, devoid of elegance and ease. In both cases the patient seemingly must recover his knowledge of using his limbs, and this may be accomplished through exercise, and suggestive therapy.

Ballet[16] believes that the cause lies in a fixed idea, the patient conceiving that he is unable, or, as Ballet puts it, is *afraid* of being unable to walk. This fear is more characteristic for psychasthenic or neurasthenic states, and does not specifically belong to hysteria. A psychsthenic, for instance, may be afraid to cross the street (agoraphobia), and the result will be inability to walk in spite of the absence of a lesion of the central nervous system. The differentiation between functional and organic disturbance of gait is based on the fact that in the functional type there is an absence of marked rigidity, exaggerated reflexes with clonuses, Babinski, Oppenheim and other signs.

Speech disorders, such as stuttering, stammering, aphasia and aphonia, frequently occur in hysteria. The characteristic features of hysterical speech disorders are their sudden occurrence after fright or sudden emotion. Hysterical stuttering in no way differs from ordinary stuttering, while so-called hysterical aphasia is *complete*, that is, there is a complete loss of speech sounds, inability to utter a single word. The patient makes no attempt to speak, as in organic aphasia, and instead of answering questions, writes them down. In other cases, loud speech is impossible, whispering only being possible. When speech is lost, the condition is that of hysterical aphonia, which is frequently combined with aphasia. The intelligence is preserved in both hysterical aphasia and aphonia. In still other cases words can be uttered, but the accent, articulation and intonation of the speech are changed, as pointed out by Marinesco[17] in his patient. The words are stretched, disarticulated, pronounced as in singing, some words or syllables being omitted altogether or produced in an indescribable manner (paradoxic aphasia of Marinesco). Recovery from aphasic disorders may be sudden or gradual, stuttering sometimes preceding complete speech recovery.

Visual or *ocular* disorders, commonly considered as stigmas were previously mentioned (constricted field of vision, achromatopsia, dyschromatopsia). Other visual disturbances are comparatively rare. To these belong polyopia, diplopia, macropsia and micropsia.

In polyopia the objects appear multiple, or a person sees double with one eye (monocular diplopia); in diplopia he sees double with both eyes; in macropsia objects appear enlarged; in micropsia, diminished. Apparent ocular palsies (of the third and sixth nerves) may occur in children, as the result of spasmodic contractions of the muscles they supply, causing hysterical ptosis, hysterical strabismus, external ophthalmoplegia (paralysis of the eye-balls). A spasm of the sphincter of the pupil may produce loss of the light reflex, but this is exceedingly uncommon in hysteria, in fact so rare that it should be ignored, for loss of the pupillary light reflex is an essential sign of an organic brain lesion.

A very remarkable though rare visual disorder deserves discussion, namely, hysterical blindness. Like other hysterical symptoms, it occurs suddenly following some emotional shock. The patient becomes totally blind, does not perceive even the strongest light, though his eye fundi and the pupillary reactions are entirely normal. The blindness is sometimes associated with other hysterical symptoms; sometimes it is the sole manifestation of hysteria and lasts hours, weeks, months, or years, the hysterotraumatic cases being usually very obstinate. The history of a sudden onset, the normal fundi, the preserved pupillary reaction (the triad of Dieulafoy) are of great help in differentiating it from organic brain lesions (tumors, serous meningitis, encephalitis) and uremia.

Lateral hemiopia (hemianopsia) almost always indicates an organic brain lesion, though Scheffield, Charcot and Janet admit its occurrence in hysteria.

Gastro-intestinal Disorders.—Complete loss of appetite, perverted appetite (craving for uneatable things, pica hysterica), vomiting, gastric and intestinal pain, meteorism, constipation, frequently occur in hysteria. Especially common in children is the loss of appetite; they absolutely refuse any kind of food. If forced to eat, they are seized with vomiting. There is not only absence or loss of appetite, but inability to digest food. The general condition suffers sooner or later, and anemia, tuberculosis and other dangerous complications set in, causing death. When the desire for food returns, there is complete inability to retain it, the stomach being unable to digest the mildest substances. Nervous anorexia is justly considered a dangerous complication, and should be treated at once by isolation and other measures, to be described.

Genito-urinary disturbances usually take the form of ischuria, inability to urinate. Cases have been described in which the retention of urine would last for days, accompanied by vomiting and excessive elimination of urea in the vomited masses. While many cases of hysterical ischuria are undoubtedly artefacts, caused by some manipulations of the patient to hide the urination from the physician and nurses, yet cases are described (Charcot) in which this condition existed beyond doubt. In contrast to the retention of urine as a common feature, incontinence is exceedingly unusual. Other urinary disorders are polyuria, pollakiuria (frequent urination) and oliguria (rare and deficient urination).

Cutaneous lesions, such as erythema, dermatographism, ulcers, blisters, have been described repeatedly; but their occurrence is denied by the latest observers, being considered as a vasomotor reflex Babinski).

Vasomotor disorders have also been noted, such as change of color of the skin, and edema. Goldblatt speaks of a peculiar, pinkish-blue color of the face, accompanied by slight edema and swelling, as very characteristic in hysteria. The cheeks look as if they were covered with bluish powder, the discoloration increasing upon slight rubbing.

These so-called accidental symptoms are but a few of the many that may be observed in hysteria. Accidental symptoms may also occur in the respiratory organs (coughing spells which completely resemble those of whooping-cough), hiccough (spasmodic contraction of the diaphragm), secretory organs (sweating, salivation, abundant secretion of tears). They are usually associated, and this is important in diagnosis, with the peculiar manifestations which make up the character of the hysteric. An hysterical patient, like an epileptic, is eccentric, lacks the altruism which is so frequent in children. He is selfish and vain and tries to evoke interest and sympathy for himself. All his thoughts, feelings and actions are centered upon himself and his morbid sensations. The affective sphere is very much involved, reacting violently with the slightest cause. The attention is poor. It is hard for an hysterical patient to concentrate; he is, therefore, incapable of reading, studying or working. Contrary to the mental condition in epileptics, hysterical patients are intellectually keen and bright, but highly suggestible and lacking in will power (abulia).

Hysteria may be associated with any organic brain or other lesion, such as tabes dorsalis, multiple sclerosis, epilepsy (hence the term hystero-epilepsy), and it may dominate the clinical picture to such an extent as to make a proper diagnosis exceedingly difficult.

A **diagnosis** of hysteria should be made only after excluding organic brain or spinal cord lesion. The principal differential features have been previously outlined. In the presence of the stigmas (anesthesias, contracted field of vision, paralysis, etc.), a diagnosis is comparatively easy. In their absence, one can exclude hysteria if the signs are present by which it can be excluded with certainty. These are degenerative muscular atrophies, loss of tendon reflexes, the presence of Argyll Robertson pupil, the Babinski sign, optic atrophy, papilledema, cranial nerve palsies, incontinence of bladder and rectum. It is true that cases of hysteria have been recorded in which an Argyll Robertson pupil was present and the cranial nerves (third, sixth, seventh and twelfth) have been implicated; but all such cases are recorded in the older literature and need revising. A true hysteria usually lacks such symptoms and in their presence the diagnosis should not be hysteria. Of the non-nervous lesions, especially in children, hysteria should be differentiated from *coxalgia*, *Pott's disease* and *abdominal pains*. In true coxalgia (hip-joint disease) the onset is slow; spontaneous pain is absent, but pain is severe on pressure; the position of the affected limb is typical; the gluteal fold is absent, and atrophy is shown. In hysterical coxalgia, the onset is sudden, following an emotional shock; the pain is spontaneous and very severe, but absent on pressure; the skin near the joint is hyperesthetic; the position of the limb varies, and atrophies are often marked (Bézy).

In hysterical Pott's disease (pseudo Pott) the contractures of the lower limb may disappear under chloroform narcosis. The pain is due to hyperesthesia of the skin or of the vertebral column, the paralysis being of the functional variety (no Babinski sign, for instance). Great caution should be observed in diagnosing, as hysterical, abdominal pain which may simulate appendicitis or peritonitis. Aside from the clinical picture, one should consider the blood examination, temperature, course, and mode of onset.

Course and Prognosis.—Hysteria is essentially a chronic malady. Its course is more or less protracted, depending on individual features, the etiologic and psychic factors, possible accidents and attacks. For instance, if evident changes have taken place in the character make-up of the patient, the more morbid the personality, the worse the surroundings, or the more defective the heredity, the worse, accordingly, is the prognosis. Generally speaking, children may recover from the severest types of hysteria, for they are more suggestible and become victims of this disease from milder causes, which are more easily removed. In other words, the morbid states arise easily and as easily disappear. The statistics of Tobias[18] and Schmidt show that in one-half of the cases, hysteria in children can be cured completely. Besides, as in the majority of chronic diseases, there may be remissions, or one set of symptoms may be replaced by another, the clinical picture, as stated, being variable and mobile. As to life, the prognosis is generally good, except in cases of nervous anorexia (loss of appetite and vomiting), which renders the prognosis more serious.

Paralysis, contractures and anesthesias may be persistent, while sensory disturbances may also become permanent. Fortunately, such occurrences in children are rare.

Theories.—One very common theory is that hysteria is the result of acute, sudden emotions which affect the psychic state of an individual, thus causing the paralysis, speech troubles, visual disorders, pains. The patient does not or is unable to defend himself by suppressing the emotions which produce these results (Raimiste).[19] In other words, there is no inhibition, which may result from chronic emotional factors (worries, struggles for existence). Another theory is that there is loss of certain perceptions (Charcot), the paralysis, for instance, being due to the loss of images of motion, the patient forgetting how to use the limbs (hence paralysis), how to feel (hence anesthesia), or how to walk (astasia-abasia). Möbius modifies this view somewhat. He states that perceptions are not lost, but modified—they are morbid. A patient becomes paralyzed because he sees someone else paralyzed; has a tremor because he happens to see a tremor and so forth.

Sollier[20] considers that the fundamental disorders in hysteria are due to a peculiar inactivity of the cerebral cortex, which he calls vigilambulism, in contrast to somnambulism. The cortex is more or less profoundly asleep, or only half awake, and cannot perform its functions with sufficient vigor.

Janet does not go so far; he believes that various perceptions merely do not reach the higher centers, remaining submerged in the subcortical regions, in the subconsciousness. The perceptions, especially when multiple, reach the conscious sphere only partially, or only one of the many perceptions does so, and even if they do reach it, they are not properly assimilated. Janet taught that this condition is due to a constricted field of consciousness.

The function or value of the subconscious activity is especially emphasized by Freud and those of his school. According to these observers, life is controlled in equal manner by conscious and unconscious factors (complexes), which may be motor, sensory or purely psychic (affective). Of all these very numerous factors the most important is the sexual, in the form of psychic trauma. The latter, retained in the subconscious sphere, produces various abnormal states, for if totally suppressed the psychic condition is normal. All the troubles of the hysterical patient come chiefly from the involuntary reminiscences of the psychic traumata.

The latest theory is that of Babinski, according to whom the so-called hysterical phenomena result from simulation, and may disappear on suggestion, countersuggestion or persuasion; hence the name pithiatism. By properly examining the patient, he says, it is possible to demonstrate complete absence of hysterical anesthesias, motor, visual and other disorders, which are produced not by emotion but by suggestion. In war time the emotions he says were very violent and numerous, yet they did not produce hysteria, though they might have created a predisposition to it, by increasing suggestible states, and sometimes by introducing elements of autosuggestion.

The vasomotor symptoms observed in hysteria are, according to Babinski, of reflex origin, caused by disorders of the sympathetic nervous system. In general, Babinski's teaching is partly Bernheim's modified theory of autosuggestion.

Treatment of hysteria is prophylactic and curative. In cases in which its occurrence could be expected, the treatment should be prophylactic or preventive. If the surroundings are unhealthful—hysterical mother, father, sisters—it is advisable to remove the child from the family and raise it somewhere else. This is an excellent measure even for older children, for being very impressionable, children easily acquire hysterical habits by imitation or suggestibility.

In the fully developed condition, the treatment varies according to the character of the lesion, its intensity, and the surroundings of the patient. Medicinal or drug treatment is not indicated unless in acute attacks such as tremor or convulsions, when valerian or any other strong-smelling drugs (asafetida) may be given. Otherwise the treatment should be purely psychic or moral and may be combined with the so-called psychanalytic methods, as used by Freud and his school. Through psychanalysis, the underlying cause of the disorder is determined. By taking a complete history from the earliest life of the patient, one tries to determine the probable origin of the emotions or psychic injuries that might be held responsible for the diseased state. The idea ("affect") embedded in the patient's subconsciousness, torturing and distressing him, should be dug out, as it were, and brought to light, made conscious, and removed by means of persuasion, suggestion or other means. Ormerod[22] declares that relief is experienced by "giving vent to pent up mental distress."

In the case of children psychanalysis is confined to taking a good substantial history and is of course much less complicated than in adults. The psychanalysis is combined with the persuasion treatment. The patient's complaints must be thoroughly investigated, his confidence should be gained, and authority exercised by the physician, not only over the child but also over the parents and relatives. In other words, there must be a full coöperation between the patient, his parents, and the physician. When it is impossible to obtain such cooperation, no effective results can be gained, and the patient should be removed from his unfavorable surroundings and placed under the personal observation of the physician. Sollier brought out the benefits and advantages of such treatment excellently. First, it is easier to control the patient; second, it is possible to study and observe his habits, character and conduct; third, he can be trained in his eating habits, rest, bowel regulations, sleep and exercises. To accomplish this, the isolation must be complete, and absolute. No visits, letters or other communication with the patient should be permitted.

Aside from isolation, rest in bed is urgently indicated, especially when the general condition is run down, when there is great asthenia, nervous vomiting, and aversion to food. Rest is to be taken with plenty of fresh air, light, and mild passive exercises (massage, baths). Electricity can be used as a suggestive remedy.

Psychic treatment consists of talks to the patient, directed toward relieving him of anything that might prey on his mind. Moral treatment, as outlined by Sollier, consists in teaching the patient good habits, self-control, in short, attention should be paid to the smallest details of the physical and psychic spheres of the patient.

Suggestive treatment for the purpose of reconstructing the patient's personality, "awaken his consciousness," is powerful in its remedial results. It should be employed in the waking state, so to speak, *indirectly*. One may suggest to the patient the ability to feel or to walk by explaining such a possibility to a third party, or to the patient himself, or by promising him a complete cure through electrical treatment, massage or hydrotherapy. In some cases, persuasion, explaining to the patient the nature of his symptoms, and convincing him that the condition is curable is also an excellent remedy. Hypnotism is generally not advisable, especially when it is difficult to induce. In case it must be done for diagnostic purposes, for instance, or for the purpose of ascertaining the patient's complex, the nature of his psychic trauma, it should be performed with the patient's consent and in the presence of a third person.

The treatment of hysterical *attacks* calls first of all for immediate removal of everybody around the patient. Quiet should be created first, this alone being sometimes sufficient to stop the attack. In many cases, pressure on the hysterogenic zones (ovarial region, for instance), stops the seizure. In general, treatment of hysteria in children is easier than in adults, with their fixed ideas, changed personality, disagreeable or unfortunate experiences in life, obstinate, long-standing contractures and paralyses with anesthesias, all of which symptoms in children are usually mild and, therefore, easier to control.

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TICS

Definition.—Tic is a motor restlessness which manifests itself in peculiar muscular contractions. The latter are, as a rule, clonic, i.e., brisk, sudden, abrupt, momentary movements. In some cases, as in the tics of the neck (mental torticollis), the muscular contractions may be tonic—somewhat prolonged, even tetaniform. The contractions in a given form of tic are always of the same character, always reproduced in the same fashion and confined to the same muscle or group of muscles, resembling in their sum total an automatic though physiologic act. But the tic is performed without motive, cause or reason, is ill-timed, as it were, being superfluous and altogether uncalled for. In other words, the act, though coördinated and logical, is purposeless.

Another feature is that the patient is unaware that he is performing a physiologic act which is not only purposeless but also irresistible, prone to repetition. The impulse to repetition can often be repressed for a short time, provided the patient's will power is strong enough. Ordinarily, it is not sufficiently strong to resist the impulse, the gratification of which is usually accompanied by a feeling of satisfaction, while its suppression is associated with a feeling of dissatisfaction. The muscular contractions are, as a rule, painless and do not, therefore, interfere with the patients' daily movements, actions or work. For instance, Guinon's[1] patient, a famous ballet dancer, could perform the most marvelous dancing feats without showing a single tic, but after he was through with the dancing he had an abundance of them. The tic movements can be repeated or reproduced by the patient at will; they are not continuous, but follow one another at regular intervals, and may subside totally in the presence of strangers, becoming abundant when the tic victim is alone. During sleep they always cease.

Varieties of Tic.—A tic may appear in any muscle of the body, or, which is more frequent, in a group of muscles. For instance, the deltoid alone may be involved, causing shrugging of the shoulder, or the muscle closing the eye may be involved, producing blinking tic; likewise the muscles of the face, neck, nose, lips, mastication, upper and lower limbs, toes (Raymond and Janet[2]), and even of the bladder (Oppenheim[3])—all may cause specific tics. Hence, there are recognized facial tics, neck tics, lip tics, etc. By far the most frequent variety in children is the facial tic, while the neck tic occurs in much older persons. One individual may show several varieties, a facial tic, for instance, interchanging with a shoulder or neck tic, and *vice versa*, or may be co-existent with other types, that is, tics may occur in combination.

Facial tic is a sudden rapid contraction of any muscle of the face, as of the forehead, cheek or nose. The *eyelid tic* is a sudden clonic contraction of the eyelid muscles, causing closure of the eye. *Mastication tic* consists in lowering and raising of the jaws, chewing movements, and so forth. The resemblance of a tic contraction to a spasm gave rise to the names convulsive, or spasmodic, tic, these names having been considered synonymous. Through the work of Brissaud,[4] Meige and Feindel,[5] Nougués,[6] Babinski,[7] Patrick[8] and others, it became possible to establish the typical features by which a spasm, in the face, for instance, could be differentiated from a convulsive tic, and at the present it is definitely recognized that a facial or any other spasm differs totally from a tic, these being entirely different clinical entities. In a facial spasm, the contraction, though sudden, resembles more the one produced by an electric current applied to the trunk of the seventh nerve, the contraction being partly tetaniform, partly clonic and jerky, accompanied by fascicular quivering which may begin or end the spasm, while nothing resembling this occurs in tic. The former is mostly unilateral, being the result of some lesion of the reflex arc (Babinski), while tic does not signify any neuronic lesion, being a purely psychic or psychomotor phenomenon. Irritation of the conjunctiva by a speck of dust or any foreign body, will cause a spasm of the eye muscle which will persist until the speck is removed. The muscular contractions will, in such a case, be perfectly legitimate acts. When, however, the speck is gone and the contractions of the eyelids persist, causing frequent blinking, we have a tic, for there is no occasion for the unnecessary contractions, their original cause having disappeared. It follows that a spasm may, under certain conditions, lead to a tic. The former may be painful, according to the cause; the latter is usually painless, the patient being unaware of the contractions. Spasm may interfere with the voluntary physiologic movements; tic does not. The former persists in sleep; tic disappears. Signs of peculiar mental condition (pyschasthenic states) are absent in spasm; they are usually present in tic. In spasm of the facial muscles there is slight disfiguration of some parts of the face, such as deviation of the nose, which is absent in tic. In short, spasm is an *organic* functional nerve disorder; tic is an *inorganic* functional nerve disorder. The same considerations may help in differentiating tics and spasms occurring in any other part of the body, for instance, in the neck muscles (spasmodic or mental torticollis).

In children this type of tic is uncommon, the youngest patient being that of Raymond and Janet (14 years old). In mental torticollis, the head suddenly turns toward one side, the chin to the opposite, the shoulder being raised on the side of the inclined head where the sternomastoid muscle becomes contracted and tense. The frequent contractions of this muscle may result in its hypertrophy, caused by hyperactivity, while the fellow muscle of the opposite side becomes atrophied. Although generally clonic, the muscular contractions in neck torticollis are sometimes tonic. The result of the tonic contraction of the sternomastoid is a rather prolonged deviation of the head. Any active attempt to reduce the rotated head to a normal position is futile, however strong the physical effort at reduction may be. However, a mere application of the patient's finger or hand to the inclined head or chin may stop the torsion of the head at once (Brissaud, Meige and Feindel, Patrick).

Like the facial tics the neck tic or mental torticollis is the result of some previous, often repeated purposeful and deliberate movements, which ultimately become purposeless and impulsive. L. Pierce Clark's[9] patient, a neuropath, would frequently turn his head to one side while working at his desk, as he had been told he might be stabbed in the back. This perfectly deliberate movement ultimately developed into a neck tic, since the patient kept on turning and rotating his head long after the supposed danger to his back disappeared. Another patient of Pierce Clark, a girl, would turn her head upward while taking instructions from a tall person. This upward movement developed into an impulse to turn the head upward when the occasion for such movements passed. These cases are examples of so-called psychic origin of a neck tic. There may be purely organic causes, for instance, constant twitchings of the head to get rid of a disagreeable sensation (caused, for example, by a tight collar). This may result in purposeless rotations of the head and lead to a tic.

As in the case of facial tic, *spasm* of the neck muscles must be differentiated from tic. In children, especially, so-called spasmodic contractions of the neck are unusually frequent, leading to involuntary, rhythmic movements of the head, its rotation, nodding, negation movements, and known as nodding spasm, spasmus nutans, gyrospasm, salaam tic, etc. This usually occurs in the first two years of the child's life, even as early as three months. The child moves the head anteroposteriorly, or which is more frequent, laterally, at the rate of from 60 to 80 (Abt[10]) nods a minute, or less. The movements are interrupted by a pause of some length, followed by another spell of nods. The nodding is often accompanied by slight flexion of the trunk and nystagmus, which may be limited to one eye and a tendency, as Still[11] pointed out, to look out of the "corner of the eyes, with the head slightly averted and the face turned slightly downward." When the child's head is supported the nodding stops, but reappears when unsupported. The sideway gaze and the nystagmus are looked upon by Still as almost pathognomonic for the spasmus nutans. The nystagmus is rapid and fine and may precede by weeks or months the head movements, or may exist without the latter (Still), and disappears entirely after a few weeks or months. The causes of this spasm are ascribed by some authors to rickets, dentition, ear syringing, or to some other peripheral irritation, while Féré[12] considers it as epilepsy and Raudnitz thought that ill-lighted dwellings were the cause. Whatever the cause, the prognosis is good, the malady disappearing within from three to four months (Abt).

The *differential diagnosis* must also be made between this condition and the stereotyped movements known as nocturnal rolling of the head (jactatio capitis nocturna). This occurs only in children just past infancy, when in a recumbent position, being rather a *voluntary movement unaccompanied by nystagmus* or the "*gaze look*" of Still.

To the same class of stereotyped movements belongs the so-called "balance tic" of Cruchet:[13] there are rotary movements of the upper part of the body, only during sleep, as many as from 80 to 90 movements occurring a minute. In one case, a three year old child, Cruchet observed his "tic de balancement" also in the waking state.

The foregoing types of stereotyped movements differ from tics and spasms in the absence of the brusque, spasm-like movements, of impulsive features and of free intervals. In other words, the so-called nodding spasm, the nocturnal rolling of the head and the "balancing" tic are neither spasms nor tics, but mere stereotyped acts (stereotypies).

The facial tic and the stereotyped movements are most frequent in children, other types of tics or similar movements being rare, and, therefore, they will only be mentioned. These are *shoulder* tic, sudden shrugging and raising of the shoulder; *lip* or lick tic (cheilophagia); *tongue* tic, sudden protrusion of the tongue; *arm* and *hand* tic, various movements of flexion, extension, pronation and extension of the upper limb; *gait* tic, or tic of genuflexion, when various unprovoked and useless movements are performed during walking, such as flexion of the knee, striking the ankles against each other, turning several times around the axis of the body and striking the back with the heel of the foot; *foot* and *toe* tic, as reported by Raymond and Janet,[2] in which extension of the great toe and abduction of the neighboring toes caused a varus position of the foot during walking.

Further, there should be mentioned *larynx* tics (sudden emission of noises, syllables or words), *bladder* tics to which Oppenheim refers the diurnal bladder and urine incontinence, due evidently to sudden contractions of the abdominal musculature, and so-called tics of ideas (tic de pensée, Chabbert[14]). In the latter type of tic the patient, having posted a letter, for instance, impulsively returns many times to the mail box to ascertain whether the letter has been dropped in the box; or it occurs in form of repeated counting of pennies or windows, for instance.

Some of these tics, as tic de pensée, laryngeal tics, and gait tics are but symptoms of so-called generalized tics or tic malady (Gilles de la Tourette's disease). Thus, we must admit two large groups of tics. In one group, the tic is strictly limited, as eyelid tic; in the other, the tics are generalized, spread over several portions of the body, simultaneously affecting the face, shoulders, larynx, etc.

The clinical picture of general tic consists of a motor *organic* element, combined with mental or psychic features. The organic manifestations comprise various involuntary movements which are first localized, in the form, for instance, of twitchings in the face, around the mouth, etc. Later on the movements become generalized, involving the shoulders and the muscles of the body. The child raises the shoulders, turns the body to the right or left, bends forward or backward, flexes the legs, jumps, emits inarticulate or articulate sounds, and so forth. For a long time the involuntary movements may be the sole manifestation of the tic malady. Sooner or later, they become complicated by mental symptoms which Gilles de la Tourette[15] described as echolalia, coprolalia, echokinesis (echopraxis), and to which Guinon added pure psychasthenic signs. Echolalia is a repetition of sounds or words heard weeks or months previously, even when spoken in a strange language. The word repetition is, like the muscular tic contractions, sudden and abrupt, and is compared by Guinon to a muscle twitch. The sounds may be meaningless, like hem, hem, ouh, ah, or they may appear as articulated words. Sometimes they are of obscene character (cursing, swearing, irrespective of time and place) and Gilles de la Tourette, names this coprolalia. Imitation (voluntary or impulsive) of actions is termed echokinesis, or "echolalia of acts and gestures" (Gilles de la Tourette). The psychic symptoms comprise fixed ideas, bizarre actions, such as inability to open the door without turning the handle several times, various fears (of closed space, for instance), repeatedly looking under the bed before retiring, or senseless counting (arithmomania), etc. One of Charcot's[16] patients would emit, in his presence, peculiar crying sounds which seemed to express pain. The patient's mother informed Charcot that the child's exclamations were exactly similar to those which she herself emitted in the patient's presence after she pricked her finger while sewing. In short, the mental picture or psychic symptoms can be characterized as obsessive or impulsive acts, or, as Charcot put it, as tic of ideas. The latter, in combination with the motor tics, make up the clinical picture of tic malady or general tic (Gilles de la Tourette). In some cases it may be confined to one symptom only, as echolalia or generalized twitchings. It does not occur in very young children, not before the age of six, and mostly in those with defective heredity. Yet the patients themselves are not mentally defective; on the contrary, intellectually they may be even superior to an average child. Charcot named them unbalanced (déséquilibrés). In the presence of psychic symptoms and defective heredity, the tic malady differs from minor or Sydenham's chorea with which it is generally confounded, for the psychic manifestations—echolalia, coprolalia—may be absent for a long time. Yet, even in the sole presence of the twitchings, it is possible to differentiate them from chorea. In tics, the movements, generalized as they may be, are always sudden, brisk and coördinated, while chorea movements are comparatively slow and never coördinated. The tics are interrupted by regular, more or less prolonged intervals, which are absent in chorea. The muscle contractions in tics are always the same, preserving their individuality and resembling a definite physiologic act. In chorea the movements vary *ad infinitum* in their type and character, changing constantly. They are irregular, arrhythmic, and illogical, and cannot be restrained even for a moment. Chorea movements continue in the presence of strangers, increasing when observed, while the tic performances are preferably carried on in seclusion, rather subsiding when observed, and can be reproduced at will, which is not the case in chorea. There are some cases of chorea in which the various polymorphous generalized twitchings are combined with mental symptoms like those observed in generalized tic. They form, as it were, a transition from chorea to tic and have been described by Brissaud as variable chorea. When the various generalized movements are more constant in type, more stable, systematized and coördinated, than those observed in Brissaud's variable chorea, they are called variable tic (Meige[17]). Speaking of this type of tics, Meige says that they, like any other tic, reflect the victim's mental state, being its product. If the victim has fixed ideas, his tics will be fixed and localized; if inclined to obsessions, the tics will have the stamp of obsessions. If fickle, irritable and changeable, the tics will be of the same character, mobile and variable, appearing in various definite parts of the body, succeeding one another without definite plan or order. Yet they are tics, for they always reappear in the same fashion, as coördinated, systematized movements. Each has its causes, its purpose, but the causes are fleeting, multiple, and variable (Meige).

There are many other states in which involuntary muscle contractions, localized or generalized, are observed. I may mention among others: (1) Hysteria, in which the muscular contractions are more or less rhythmic and stereotyped, resembling, for instance, hand movements in hammering (malleatory chorea); (2) athetosis; (3) myoclonus or paramyoclonus; (4) myotonic contractions and many other forms which, so far, have not been well studied or defined. One type I believe deserves a cursory mention, the so-called dystonia musculorum progressiva or deformans. This is a type of motor restlessness in which there is a torsion spasm of the trunk and extremities associated with chorea-like, or other more or less sudden, muscular contractions. They subside during rest and cease during sleep; but they increase from emotional stimuli or muscular activity, such as standing or walking and involve, as a rule, the musculature of the trunk and the lower extremities. The muscles of the face and articulation are spared, those of the neck may be involved in the terminal stage.

Nature, Prognosis and Course.—Tic is essentially a psychic disorder, occurring in persons with defective heredity. No anatomic changes of any kind have been found so far. The main physiologic feature is impairment of the volitional element; there is volitional debility. The latter may be the result of imitation, or it may be a defense mechanism against some unpleasant physical or psychic irritation, or it may be caused by an idea. Solomon[18] looks upon tics from the biologic point of view, considering them as "reactive response to some unusual mental strain," or "a constitutional, biologic, defense reaction, an organic means of adaptation." L. P. Clark[9] tried to explain tics from the psychologic point of view, following Freudian teachings, namely, that the essence of habit movements is an auto-pleasurable act based upon an inherent defect. When the auto-pleasurable acts are inhibited or modified, they continue in tic movements.

Whether the biologic, psychanalytic, or ordinary common sense explanations be given to the nature of tics in general, the fact remains that in each case the causes must vary, and the prognosis as well as the course and treatment will be different. Thus, in ordinary localized tics, the prognosis is fairly good; in generalized tics, especially of the form of Gilles de la Tourette, it is less good, though even they may recover (Oppenheim).

The course is chronic and painless, without incapacitating the patient to any great extent.

Treatment.—As laid down by Brissaud, Meige[19] and Feindel,[20] it consists of absolute rest, hydrotherapy, regulation of habits, gymnastic exercises and suggestion. Psychic discipline must be combined with the psychomotor discipline of the muscles (Meige, Brissaud). It has for its purpose the suppression of automatic acts, and their replacement by appropriate and regular acts. The patient and educator must coöperate; the former is taught the habit of exercising over his acts a control which he lacks. This control Meige accomplishes with the help of a mirror, and he asserts that tics so treated may disappear entirely. Brissaud's motor discipline is in teaching the patient to preserve immobility by performing appropriate exercises. Other methods are directed toward rendering the pleasurable movements disagreeable, by holding, for instance, a sharp instrument before the patient's face in case of facial tics; some use the psychanalytic method, or other procedures. Any method may remove the symptoms or effect a cure in mild cases. In severe or complicated cases, in which many tics are combined, a complete cure can hardly be expected, for all depends upon the mental state of the patient, which needs, so to speak, a complete reconstruction in order to make him coöperate. This is by no means an easy task; in fact, it is hardly possible. Medicinal treatment, bromids and arsenic, are altogether useless; likewise is surgical treatment (of mental torticollis, especially). Though it was successfully used in some cases (Annandale[21]), it is at present abolished.

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HEADACHES AND MIGRAINE

It is generally recognized that a headache is not a disease *per se*, but a mere symptom of some organic or functional nerve lesion. Therefore, in these conditions, a great number of possible etiologic factors must be borne in mind. They will be given only a general review here. Features of headaches specific in children, however, will be discussed in some detail.

Class 1.—Organic brain diseases (tumors, abscesses, syphilis, vascular disorders, acute inflammation of the brain substance—encephalitis—or its membranes—meningitis) are usually associated with headaches, more or less persistent, diffuse or local, often combined with vomiting, visual disorders, disc changes and other symptoms. Generally speaking, chronic headache is one of the most prominent so-called general symptoms of brain tumor. Of practically the same character is the headache in brain abscess, while in syphilis, the headaches are likely to show evening exacerbations. In encephalitis as well as in meningitis (purulent, cerebrospinal, tuberculous, serous) they are acute in *onset* and *course*. The diagnosis of this group of headaches is made from the history and concomitant symptoms, especially from serologic examination of the blood and spinal fluid. Of vascular cerebral disorders (hyperemia, anemia, hemorrhages, thrombosis, embolism), the most important in children is anemia of the brain, which may be a partial manifestation of leukemia, pernicious anemia and chlorosis. The examination of the blood is of great value in the diagnosis of such cases.

Class 2.—In a great number of cases in Class 2, the cause of the headache is not in the brain itself, but in more or less remote organs. Some of these headaches are designated as *reflex*.[1] Thus, diseases of the nose, ears, throat, neck muscles, teeth, sinuses of the head, eyes, kidneys, stomach and intestines may cause an obstinate, chronic headache, which can be remedied only by treatment of the organ at fault. Especially obstinate are the so-called sinus headaches, and the headaches due to the closure of the frontal sinus without suppuration. On account of their paroxysmal course, they may be mistaken for neuralgia or migraine.

Another equally obstinate group are the so-called muscular, rheumatic or indurative headaches, which are supposedly caused by indurations or nodules occurring in some muscles, as the trapezius, scaleni, sternocleidomastoid, at their points of insertion in the neck. The occurrence of such a headache is quite rare in persons under the age of 20, is usually occipital, but may radiate from the occiput to the frontal or temporal areas and to the vertex. Patrick[2] points out that acute cases present some elevation of temperature and a mild leukocytosis. Transient or constant pain also occurs on movement of the neck, especially extreme rotation or lateral flexion. This probably comes from the involvement of the joints, or muscles and ligaments of the neck (Patrick). The majority of authors lay much stress on the presence of nodules and small lumps at points of insertion of the muscles on the occiput, which may be detected by the finger well covered with petrolatum or any other oily substance Patrick recommends ignoring the nodules and lumps clinically, as the tenderness suffices for diagnosis. The muscular headache is most probably infectious in origin, coming from the tonsils, teeth, joints, etc. The treatment of these primary foci of infection, combined with symptomatic therapy (heat, pyramidon, salicylates), usually gives excellent results.

Of ocular lesions, refractive errors, especially glaucoma, should be mentioned, as in this the headache is frequently mistaken for trifacial neuralgia. The frequency of ocular headaches due to refractive errors is probably exaggerated, but their importance is great, for they yield easily to treatment.

It must always be remembered that headache may be caused by subacute or chronic kidney lesion (various forms of nephritis), when headache is a partial indication of uremia. It is necessary, therefore, to examine the urine in every case of headache, so as to exclude, if possible, this danger.

Gastro-intestinal disorders (chronic constipation, dietetic errors, tapeworms) cause so-called "dietetic" headaches, which can be alleviated by proper dietetic treatment.

Class 3.—A large group of headaches, acute in onset and course, occur in acute infectious diseases (typhoid, malaria, erysipelas, etc.) and intoxications (alcoholism, nicotine or ptomain poisoning). In all these diseases, the headache is overshadowed by other clinical symptoms, which facilitate the diagnosis.

Class 4.—To this class belong so-called functional headaches, which occur in functional nervous diseases—neurasthenia, hysteria, hypochondriasis and so-called "growing" headaches.

In neurasthenia the sensation is not so much one of headache as pressure, diffuse in character and covering the entire skull cap, with the feeling of constriction similar to that caused by a tight cap. On the contrary, in hysteria the headache is local rather, confined to a small spot, and is commonly called clavus, as the pain resembles the driving in of a nail.

The headache in so-called hypochondriasis is psychical, having no specific features, and is overshadowed by numerous somatic difficulties. In children, this type of headache is generally of small importance, as hypochondriasis is an unusual occurrence in youth. Of much greater significance and interest in children is the so-called "growing" headache, which occurs usually from 9 to 12 years of age. The victims of "growing" headaches are poorly nourished, pale, chlorotic They grow rapidly and easily become fatigued from slight mental efforts, which cause the headache. Keller[3] says that the headache is usually frontal, seldom temporal, and never occipital. It is ordinarily severe, shooting, throbbing, tearing, not confined to one side, as in neuralgia, and independent of any eye lesion. It subsides after a good night's rest;

but becomes worse during school hours or in the evening. Hutinel and Babonneix[4] remark that children with "growing" headaches usually have a very poor appetite, complain of pains in various parts of the body, and are sad and hysterical. Keller justly points out that such headaches in children are a source of great anxiety to the family physician and parents, as meningitis or some organic brain trouble may be feared. Rest, fresh air, change of environment, proper dietetic measures, regulation of the bowels, and hydrotherapy are excellent remedies for "growing" headaches, which may be considered constitutional, as a partial manifestation of changes going on in the growing human organism.

Class 5.—Both migraine and nasal headaches are syndromes in which the headache dominates the entire clinical picture and stamps both as distinctly specific clinical entities.

Migraine, hemicrania, or unilateral headache occurs frequently in children. It is a symptom-complex in which the headache is peculiar, specific, mostly unilateral and occurs in paroxysms. It is sometimes preceded by an aura, and is often followed by gastric disturbances (nausea and vomiting).

Heredity is most important in considering the causes of migraine. It may be direct (migraine in the parents), or indirect (various other nervous and mental disorders in the family). Heredity is so constant and prominent a factor that Möbius[5] and Filatov[6] assert that they hesitate to diagnose migraine in cases in which the hereditary factor is absent. Another factor is the age—young people and children being most frequently predisposed to this malady. In fact, many cases of migraine in adults can be traced to a beginning in early childhood. Flatau,[7] in a history of 500 cases, shows that in 12 per cent. the trouble started before the age of 15, while cases arising late in life (after 50, for instance) are not migraine, but are due to some organic brain lesion. Migraine is not a product either of heredity or of the disturbances of adolescence alone. There must be some exciting cause before the latent migraine becomes manifest. Various intoxications (alcohol, nicotin), infectious diseases, irregular living habits (mental stress, sexual irregularities), diseases of the eyes, throat, sinuses, may induce it. In short, the existing equilibrium may be upset by any of these causes. In the presence of strong hereditary predisposition, an attack of migraine will occur with slight provocation. If the predisposition is mild, an attack is caused only by grave provocation. In some cases, the attacks are, so to speak, spontaneous, that is, they occur without any apparent cause. Charcot,[8] Féré[9] and Oppenheim,[10] have observed typical migraine in tabes, paralytic dementia and cerebral thrombosis. It is, therefore, called symptomatic in contrast to the idiopathic form in which no organic brain lesion can be demonstrated.

A typical attack of migraine consists of (1) a *prodromal* stage; (2) *aura;* (3) the *attack* and (4) *post-migrainous* symptoms. In the prodromal stage there is lassitude, disinclination to physical or mental work, gastric, urinary and visual disturbances, vertigo; or the patient is depressed and irritable. In some cases this stage is absent, or, when present, is followed after a few hours, by an aura or premonitory symptoms. Various subjective sensations immediately precede the headache, such as numbness, paresthesias (formications in the hands, feet, lips, tongue), visual, auditory, and psychical manifestations. Some of these, such as the formications or tingling, ascending from the hand to the arm, much resemble the aura in epilepsy.

However, as Gowers[11] remarks, it never begins in the foot and may last 20 minutes, while in epilepsy, but a few seconds elapse before the attack begins.

The visual auras, such as ophthalmic manifestations, which sometimes obscure other signs and symptoms, occur more frequently and have been more closely observed. Galezowski,[12] calls this form "ophthalmic" migraine. He groups the ocular manifestations as: (1) *periodic hemiopia;* (2) *scintillating scotoma;* (3) *amaurosis migrainosa* and (4) *photophobia migrainosa.* In hemiopia, the patient sees but one-half of the visual field, one side of a person's body, one-half of the letter while reading, etc. In scintillating scotoma the patient sees luminous, bright bodies, round, irregular, angular or zigzag in shape, or rapidly turning, moving, revolving and dancing circles, or rainbows, "ancient fortifications," etc. A dark spot appears in the center of the luminous bodies and, gradually enlarging, obscures the visual field partially or entirely. Often a dark spot is seen without the illuminating contours. This is called central scotoma. In other cases a cloud appears to cover the field of vision. Galezowski remarks that the scotoma may last several hours, days, or months, disappearing and recurring, but preserving the same shape and character in each recurring attack.

Amaurosis, or complete blindness, also described as "blind" headache, disappears when headache and vomiting set in.

In photophobia or extreme sensitiveness to light rays, the patient craves darkness. Sometimes the sense of smell is also extremely keen. About half an hour after onset, the aura is followed in typical cases by an attack of headache. It is usually unilateral, though the entire head from above the eyes to the occiput may be involved, the pain radiating to the neck, shoulders and arms. The pain in the head is acute, tearing, boring, pressing and hammering, lasting hours or days. It greatly exhausts the patient, who stays in bed, with the head buried in the pillow. The face is pale or flushed. There is inability to do physical or mental work, or it is done mechanically. Lack of appetite or complete aversion to food is shown. The attack terminates gradually, often with vomiting, sometimes with sweating, polyuria, nasorrhea, and is often followed by sleep. In children the temperature may rise during the attack, which may last for 24 hours, reappearing after several months. According to Gowers, such recurrent feverish attacks may be replaced in later childhood by typical headaches, and are looked upon as equivalents of migraine. In Bary's[13] case the attack was characterized by gastric pain. The patient would be seized with sudden pains in the stomach having no relation to food, and disappearing after one or two hours of sleep. These attacks persisted for three months. During this period no headache appeared, but with the disappearance of the gastric pain headache began.

In Charcot' scases,[14] speech disorders (motor or sensory aphasia) complicated the attacks. These may last for hours, rarely for weeks. In some cases these disorders alternate with epileptic seizures. Migraine and epilepsy may occur simultaneously, that is, an epileptic fit may come on during the attack of migraine, or the petit mal attacks may turn into migraine. Such occurrences have given rise to the opinion that epilepsy and migraine are identical. There are resemblances, such as the premonitory symptoms, aura, and the attack followed by sleep; yet dissimilar features are not wanting. In migraine the phenomena are sensory; in epilepsy, they are motor (convulsions). In migraine the premonitory stage and the aura are prolonged; in epilepsy they are brief. In migraine the clonic spasm is unknown; in epilepsy, Gowers declares, the so-called tingling zone, passing slowly from the hand to the arm, does not appear. In migraine the delirium, somnolence, stupor and loss of consciousness observed occasionally result from pain; in epilepsy they follow the epileptic fit. Gowers, Hubbell[15] and many others deny that these two disorders are related.

During or before attacks of migraine in children no psychical disturbances, such as illusions, hallucinations, psychomotor restlessness, and dreaming states, have been recorded. Equally uncommon is the complication of third nerve paralysis (ophthalmoplegic migraine), though Flatau records such cases in a child of three and one of six years. In the latter, migraine began at the age of 11 months. When it does occur, third nerve paralysis *follows* the headache, attacks the same side, and persists for days or months. The paralysis may be partial, or may involve the fourth and sixth nerves. When total, it involves all the, extrinsic and intrinsic eye muscles. After a few days or months, the ophthalmoplegia disappears entirely or partially, although it may recur after a short or long interval.

Generally speaking, migraine in children is milder than in adults. Fabre[16] says that in small children, especially in infants, the predominant symptoms are periodical gastric disturbances—severe pain in the stomach, vomiting, coated tongue, slight fever, and a mild headache. In children the periodicity of the vomiting and headache, with intervals of complete freedom, are the sole diagnostic signs of migraine. Filatov[17] mentions the pallor of the face, languor, and yawning. If these symptoms, he says, occur periodically in an otherwise healthy child, each paroxysm terminating with vomiting and sleep, migraine can be recognized even in children that cannot talk.

The prognosis of migraine in children is good as to life, but not very favorable as to complete recovery, for sometimes epilepsy supervenes, or the migraine may be a manifestation of some organic brain lesion.

Many theories, or rather hypotheses, have been advanced regarding the nature of migraine. I will mention but a few: the toxic, the reflex, DuBois Reymond's[18] vasoconstriction, and Mölendorf's[19] vasodilatation theories. The relaxation or spasm of the cerebral vessels, with resulting paralysis or irritation of the sympathetic nerve (narrowing of the palpebral fissure, myosis, retraction of the bulbus, congestion of the face) so often described in migraine, result from, rather than cause, the factors in the attack. Beyond question the cause of migraine is some cortical lesion, probably secondary to sinus trouble, but the essential quality of this lesion, the mechanism of the cortical perturbations leading to an attack, is totally unknown.

Nasal Headaches.—According to Sluder,[20] some nasal lesions give rise to severe headaches lacking the gross symptoms or signs that betray the nasal origin of the primary lesion. Sluder refers to (1) closure of the frontal sinus without suppuration; (2) the syndrome of nasal ganglion neurosis; and (3) the picture of hyperplastic sphenoiditis. In the first group, the headache, usually frontal, is made worse by use of the eyes, especially for near work, and is not relieved by glasses ("asthenopia"). It is accompanied by a tender point in the upper inner angle of the orbit (Ewing's sign). This sign is not found in the headaches caused, for instance, by ethmoidal or sphenoidal sinus, ocular, digestive, and other disturbances, except empyema of the frontal sinus. To remove the middle turbinate is sometimes the only measure that will relieve the pain in the head, although anything will be of service that will diminish the swelling of the soft tissues which close the inlet to the sinus. Various astringents may be applied to, in or about the middle meatus.

The second group comprises the syndrome of nasal ganglion (sphenopalatine or Meckel's ganglion) neurosis, or Vidian neuralgia, described by Sluder[21] in 1908. The ganglion is situated in the upper portion of the sphenomaxillary fossa in the proximity of the sphenopalatine foramen just beneath the maxillary branch of the trigeminal nerve. There may be a history of mild coryza, followed later by pain at the root of the nose, in or about the eye, in the upper jaw and teeth, sometimes in the lower jaw and teeth, and extending backward to the ear, causing earache; emphasized at the mastoid, but always severest at a point 5 cm. back of it. Reaching backward by way of the occiput and neck, it may extend to the shoulder blade and shoulder (less often to the axilla and breast, and in severe attacks to the arm, forearm, hand and even the fingers tips). This painful syndrome is sometimes supplemented by the sympathetic syndrome—vasomoter and secretory phenomena, which are sometimes the only symptoms of the disturbance of the nasal ganglion. These are severe, protracted sneezing, with abundant secretion of mucus, swelling and redness of the external nose, greatly reddened eyes, bathed in tears, dilated pupils, wide-open eyelids, photophobia ("terrible cold," as patients put it). Local treatment—application of cocain in or around the sphenopalatine ganglion, injections of alcohol, phenol, or formaldehyd—is very effective.

Third Group.—To this belongs hyperplastic sphenoiditis, which, in Sluder's opinion, underlies many of the recurrent headaches in otherwise healthy people who suffer from migraine. Inflammation occurs in the sphenoid bone, in which is located the sphenoidal sinus separated by thin bone plates from the second, third, fourth, fifth and sixth cranial nerves. Aside from migraine, many other obscure morbid conditions are caused by hyperplastic sphenoiditis, such as optic nerve changes, visual disorders, maxillary neuralgia, and Vidian neuralgia, as mentioned under Group 2. Sluder saw extremely obstinate cases of migraine relieved by a sphenoid operation. Some of his cases were of 40 years' duration, and many of them were observed in early childhood, as early as the fourth year.

These three syndromes, so ably studied by Sluder, should be borne in mind in obscure and obstinate cases of headaches and migraine. At any rate, they fully deserve further study, for which the remarkable monograph of Sluder[22] cannot be recommended too strongly.

Treatment of Headaches.—This must be prophylactic and symptomatic. The general physical and psychical conditions of the child must be improved by proper diet, bowel regulation, sleep, and rest. Overstudy in school and at home, late hours, and defects in the general constitution of the child, such as excessive irritability, excitability, masturbation, and so forth, must be looked for and corrected in the treatment of headaches in general and migraine in particular. The removal of possible underlying causes (in eyes, stomach, bowels, etc.), should be attempted, in order to prevent the possible occurrence or recurrence of the headache or attacks of migraine. Some authors report good results from correcting chewing or eating habits,[22] some from proper glasses,[23] some through treating sinus and nasal troubles. All these measures are in many cases strongly indicated.

A severe attack of headache, especially migraine, can be controlled or relieved by rest, hot applications, pyramidon, bromids with or without caffein, or cannabis indica, although sometimes even morphin injections may be necessary.

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DISTURBANCES OF SLEEP IN CHILDREN

Sleep is a physiologic necessity without which life is absolutely impossible. It is needed for renovation of the system, the used up energy being restored to the tissues of the body during sleep. The waste products resulting from the activity of the cells are eliminated; a regular, periodic process of repair goes on in the brain and other organs, whereby the organism is enabled to carry on its physiologic functions. Jastrowitz[1] records that activity is greatly reduced or even suspended during sleep; that of the other organs, the lungs, heart, intestines and kidneys, being retarded. Much, of course, depends on the quality of the sleep. It may be superficial or deep; the duration (quantity) may be short or long. The depth of sleep may be judged, according to Rosenbach,[2] by the state of the reflexes. In a very deep sleep, according to Czerny,[3] the reflexes (abdominal, cremasteric, pupillary, tendon) are diminished or even totally lost, and Rosenbach remarks that an irritation, such as tickling of the soles or of the nose, must be very strong to produce a reflex action. Czerny found that sleep is most intense during the first hour, markedly decreasing in intensity during the second hour, being lightest in the fifth and sixth hours, gradually becoming deeper toward morning, when, however, it is not so strong as during the first hour. As a rule, in children, sleep is deep. Children fall asleep in spite of noises and, if awakened, they fall asleep again at once. Generally speaking, children suffer from sleep disorders less frequently than adults, the varieties of the abnormal sleep being the same in children as in adults. Thus, there occur: (1) difficulty in falling asleep; (2) interrupted sleep, and (3) restless sleep.

1. In children, difficulty in falling asleep, delayed sleep, insomnia, or agrypnia, may be caused by some physical distress, such as pain or disease, itching, hunger, improper temperature of the room and poor ventilation. A properly ventilated room and a reasonable amount of warmth are most essential. In older children purely nervous causes may result in delayed sleep. Worry over school work, overstudy and neurasthenic states frequently disturb slumber during the early hours, and when these children finally fall asleep they are not refreshed. Some children cannot fall asleep when they are accidentally deprived of some stimulus to which they have become accustomed. Some will fall asleep only when they are rocked or lulled by a song, held or carried in the arms; others when they are sucking their thumb or some other object (a nipple or a portion of the bed clothes, pillow case, etc.).

2. Interrupted sleep may result from the causes given in Group 1, or from some organic disease. In some cases, no immediate cause can be found. In this classification belong night terror (pavor nocturnus), somnambulism, and so-called hypnalgia.

Night terrors occur usually during the first hours of sleep, in children between the ages of four and six, though they have been observed in older and younger children. Thus, Rey[4] recorded them in a 17 year old girl in whom they had become milder after the age of 10. Heredity does not seem to play a prominent part, since only one of several children in the same family may suffer from night fears. The child goes to bed feeling perfectly well, and falls asleep. In about an hour or two, when sleep is deepest, he wakes with a loud cry and sits up in bed. The features are distorted and express terror. The forehead perspires; the eyes stare, are wide open, and the pupils are contracted (Steiner[5]). The child is totally unconscious, failing to recognize surroundings or to respond to questions, and cannot be soothed. The pulse is accelerated and the heart impulse increased. The attack cannot be cut short and, according to Wertheimer,[6] lasts from 5 to 50 minutes, subsiding gradually. The child, sobbing, falls asleep and does not recall the occurrence in the morning. The attacks may occur every night, once a week, or less often. In some cases several attacks occur during the same night, or the attacks are abortive, the child suddenly waking up and muttering a few unintelligible words. There is profuse sweating, especially of the forehead and hands, and the hands and feet tremble. The former is considered by some authorities (Wertheimer, for instance) a constant feature. Perfectly healthy and normal children do not suffer from night fears; only those having some constitutional defect (anemia, tuberculosis, abnormally irritable nervous system) may, owing to some cerebral irritation or emotional stimuli, suffer a terrifying dream against which they react by screaming, yelling, sobbing, and sweating. In so-called predisposed children, some accidental condition, such as diarrhea, fever, difficult teething, gastric troubles, tonsillitis and adenoids may, according to some authors, evoke the attacks. Rey, for instance, reports a number of cases cured by the removal of adenoids, which, he thinks, interfere with breathing in some children through irritation of the vagus nerve fibers, which causes a sensation of suffocation. Braun[7] considers pavor nocturnus as a manifestation of neurasthenia, of which it is but one of the symptoms.

Somnambulism, or night walking, may occur in children suffering from night fears. It is characterized by the prevalence of motor symptoms. The victim wakes from his early, usually profound sleep, leaves his bed, and wanders around the house performing more or less purposeful acts—removing things or replacing them, talking, etc. The patient is unconscious, keeps his eyes open and fixed, all the senses, except the position or muscular sense, being greatly blunted. In the morning he has no recollection of the acts. In short, somnambulism is an acted dream, or a condition in which a person "dreams and acts in his dream as if he were awake and as if all the phenomena of which he takes cognizance were real" (Todd[8]). Dreams are considered by some observers as the "fulfillment of wishes," and it frequently happens that night walkers perform, during their strange wanderings, acts that pertain to their daily experience and life. Strauch[9] relates several illustrative cases. One is that of a 10 year old boy who, during one of his night walks, entered another room, pulled out the drawers of his desk and rummaged in his school materials. His mother found him there and took him back to his bed without his waking. On another occasion at midnight, he got up, walked to the second floor, and there urinated from the porch. After that he suspended himself by both hands from the edge of the porch, and was about to let himself down a 15 ft. pole when a neighbor came up and grasped him while he was still asleep. The boy had, on previous days, expressed a desire to slide down this pole, but was dissuaded.

Some authors speak of still more complex and dangerous performances, such as climbing on the roof, traversing narrow planks, the victims being utterly oblivious of danger, and their movements being carried out with remarkable precision and dexterity. The majority of authors doubt the occurrence of such complicated acts, which, however, in adults are probably of hysterical nature. Some authors, as Aschaffenburg,[10] look upon both somnambulism and night terrors as a modified epilepsy.

Hypnalgia, literally, means pain occurring in sleep. A person, for example, feels very well while awake, or before falling asleep. But as soon as sleep comes he may experience violent pain—in the stomach, cardiac region, chest, head, etc. The pain disappears after the patient is aroused, but he experiences another attack as soon as he falls asleep again. Oppenheim thinks that "the pains brought on by sleep are hallucinations (of pain); probably they are hysterical or some other functional manifestation." In children, they have been described by Hamburger[12] and Strauch.[9] Hamburger believes them to be the equivalents of night terrors, and to these he also refers the nocturnal incontinence of urine, calling all the sleep disorders in children "stereotyped" or "sleep habits."

3. Restless sleep comprises a group of cases in which the child exhibits motor restlessness, without waking up, without leaving the bed, and without noisy manifestations, such as screaming and moaning. In some children the whole body jerks suddenly, or short clonic muscular contractions occur in the hands or feet; some grind their teeth, sigh, or talk; small children exhibit so-called spasmodic deglutition, or deglutition spasms, which Zappert[13] thinks are caused by the falling back of the tongue. A noteworthy subvariety of restless sleep includes the so-called stereotyped coördinated movements, such as rolling the head from side to side, sucking, chewing and similar movements. In the head rolling, the movements are rhythmic, continuing throughout the night or part of the night; recurring with remarkable regularity, and remaining unchanged for many years. This condition is called by Swoboda and Zappert *jactatio capitis nocturna* (nocturnal jactation of the head). The rolling does not disturb the child's sleep, being regular, like that of the pendulum of a clock. The children in fact seem to enjoy the movements, for they try to assume a position favorable to this act. Thus, Putman and Jacobi's patient,[14] a three year old child, had these so-called "nocturnal rotatory spasms" from 18 months of age. After he had been asleep for a few hours, he would turn over on his right side, draw the right arm over his head, and place the left hand over the left ear. After assuming this position he would begin to oscillate his head on the pillow from right to left, in a perfectly rhythmic manner. At first confined to the head, the rotation successively involved the shoulders, the trunk and lasted, sometimes throughout the night.

The nocturnal rolling of the head is neither a tic nor a nodding spasm, for these conditions do not occur during sleep. Zappert refers it to stereotyped movements, which from long and repeated performance finally become automatic, as do thumb-sucking, nail-biting, etc.

Prognosis.—In all the varieties of sleep disorder the prognosis is good as to life and complete recovery, much depending, of course, on the etiology and the constitutional factors.

Treatment.—This must be prophylactic and symptomatic. The former is directed toward creating healthy surroundings (proper living conditions, with good ventilation, appropriate food and temperature, regular physical and mental habits). Mental and physical overwork, excessive excitement of the child's imagination by terrifying tales, or by talking in his presence of his night terrors should be avoided. Suggestive treatment in the form of medicines (bromids, valerian preparations, quinin) are of great service, especially quinin (from 0.15 to 0.24) in pavor nocturnus. Various procedures, such as hydrotherapy, psychotherapy, electrotherapy, are being used, though in the great majority of cases we can dispense with complicated methods of treatment, as the assurance given to the mother (or to the patient) that there is no danger in the condition usually suffices. In other cases, change of surroundings may be advisable, such as transfer to a hospital, or placing the child under the care of a responsible nurse.

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THUMB-SUCKING AND SIMILAR ACTS

Thumb-sucking is a methodical, rhythmic, but purposeless movement of the muscles of the lips and tongue. Nothing is accomplished by this act, which is performed for the sole purpose of sucking. In certain instances, the mouth is empty, while in other cases the thumb, fingers, lips, or toes are sucked, and, according to the part chosen for the act, individuals are designated as thumb-suckers, lip-suckers, toe-suckers, and so forth. Sometimes another person's hand, ear lobe or breast is utilized in the act, and not infrequently foreign objects, whether palatable or not, such as an article of clothing, a pillow slip or a sheet.

Certain children are not satisfied with mere sucking, and combine with it, or intensify it by, other equally purposeless manifestations. This type of sucking Lindner[1] calls "sucking with combination." Ambler's[2] patient, for instance, was a baby who would go to sleep holding the right ear with the right hand and with the left thumb in the mouth. Goodwillie[3] reports the case of a patient who, while sucking the thumb of the right hand, would pass the right index finger over the bridge of the nose, pressing it to the right, at the same time exerting pressure upon the cartilaginous portion of the nose with the right middle finger. In this way breathing through the nose was prevented. In other cases thumb-sucking is accompanied by pressure upon the navel, the genitalia or other parts of the body by the other hand. These "pleasurable" parts are rubbed constantly, and when the child is held in the arms he strives to select similar parts in the person holding him. It occasionally happens that a "sucker" during the "combination" sucking act goes into a state of ecstasy, in which the head trembles, shakes, or nods, and the body is bent forward, while the feet jerk. He is oblivious of his surroundings, does not answer questions, and chooses a safe, quiet place where he expects to be undisturbed, and where he frequently falls asleep. Others inflict injuries upon themselves by rubbing a mucous surface until it bleeds, or by pulling out hair (Lindner). To this type of sucker Lindner gives the name "exalted sucker." Some phases of the exaltation resemble erotic states in adults, and Lindner says that the "pleasurable" parts which a child irritates in sucking are analogous to the libidinous points of adults. The difference is that in sexual life active assistance is usually sought from another, hetero-sexual individual, and only in exceptional circumstances is it confined to the person himself (masturbation). Freud[4] goes still further in his assertion that thumb-sucking is a sexual activity, gratified not on another person but on the body of the person himself, being auto-erotic. Such auto-eroticism is the origin of infantile sexuality. An infant does not know any sexual object, the sexual aim being under the control of an erogenous zone which may be a portion of the skin or mucous membrane, in which the stimuli produce a feeling of pleasure of definite quality. The sexual aim of the infantile impulse consists of the production of gratification through excitation of a selected erogenous zone.

This habit occurs with equal frequency among boys and girls. It may be observed in early infancy, in later childhood, and may even exist in adult life. The condition is as common among breast fed infants as among those who are artificially fed. It is no more frequent in the poorer classes than in the higher strata of society. Lindner assumes that every child has an inborn tendency to be a "sucker," and becomes such under certain conditions. Thus, the habit of giving children sugar teats, or similar objects, to keep them quiet, may develop this habit.

The diagnosis of thumb-sucking may be made from the general aspect of the child even when he is not observed in the act. In suckers the skin of the thumb is often shrunken, macerated, excoriated, or hardened, and the thumb is sometimes covered with papillomatous formations (Porter and Williams[5]). The nails are cleaner and softer than in normal children. The mouth is often deformed, the upper jaw protruding or displaced to one side, the lower jaw being retracted. The teeth are displaced outward, with the upper lip pushed out (Chandler,[6] Herbst,[7] Daniels[8]). In lip-suckers the lips are characteristically closed or protruded. The foregoing deformities may result in malocclusion of the jaws, though as Kingsley[9] and Galatti[10] pointed out, even a long persistence of the habit is not always followed by deleterious effects.

Goodwillie, on the other hand, mentions deformities and stenosis of the nose and nasal catarrh as complications. Ambler reports a cystic tumor of the lower lip. Masturbation as a result of irritation of the genitals in so-called "suckers with combination" is a frequent sequel.

Regarding prognosis, it may be said that some children give up the habit of their own will, while others, especially lip-suckers, resort to it only when engaged in some mental or physical work or before sleep. The habit in some types, as lip-sucking (or empty mouth suckers), may persist throughout life, while other forms sooner or later disappear.

Treatment is primarily prophylactic. An infant should not be allowed to place his thumb or fingers in his mouth. Sugar teats, nipples, or any othe robject frequently used to quiet a restless child should not be used. Covering the fingers with quinin, pepper, salt, or rubber cots is of little avail. Adhesive plaster applied to the fingers is effective in a few cases.

Padded splints applied to the elbows and held in place by straps to prevent the child's putting his hand to his mouth have been recommended (Monks[11]). Heckard[12] succeeded in curing a child by giving him a bugle to play several times a day. Webb[13] recommends a rubber ball with an oval hole into which the child's wrist is loosely fastened. Berillon[14] recommends hypnotism.

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NAIL-BITING (ONYCHOPHAGY)

Nail-biting is an automatic, unconscious, frequently an impulsive act, a morbid inclination to mutilate the fingers by constantly chewing the nails. Some persons bite other objects, such as penholders, pencils, or anything that they get hold of, especially when their attention is absorbed or when they are under emotional stress. In some localities it occurs frequently. Thus Berillon[1] asserts that in Paris, France, one-third of the school children are subject to this habit. Some acquire this habit by imitation, others because of some hereditary or constitutional anomalies. In some cases imitation and heredity may be combined. Berillon records a case of a family of six children, all addicted to nail-biting; their father was an alcoholic. It may be combined with thumb-sucking, bed-wetting, night-walking, night terrors, phobias, and other states indicating an abnormal nervous or mental character of the victim. Frequently, physical signs of degeneracy are present—microcephaly, asymmetrical face, nystagmus, twitchings. In school, according to Berillon's observations, the nail-biters show some inferiority to other children; they are poor writers, less skilful in manual exercises, are disobedient and absent-minded. The origin of this vice is probably in the instinct of the child to put every object in the mouth. The instinct is transformed into an automatic unconscious act (Berillon).

The consequences of obstinate nail-biting show in the peculiar shape of the fingers—their terminal phalanges become rounded and thickened, the touch sense may become diminished to the extent of anesthesia. Swallowing of the particles of the nails or portions of other dirty objects may lead to gastric disorders and various infections.

Prognosis and Treatment.—Onychophagy is a very obstinate habit, hard to combat and lasting for an unusually long time. The treatment is the same as outlined for thumb-sucking. Some recommend dental measures (Didsbury and Pont[2]) which consist in rendering the teeth harmless. This can be achieved by preventing the frontal teeth from coming in contact with each other, thus rendering nail-biting impossible. Pont recommends keeping the jaws slightly apart by means of metallic or hard rubber covers applied bilaterally to the upper and lower molars, so that chewing of food is possible, while nail-biting is rendered impossible. In Pont's hands, this method gave excellent and radical results after several months of treatment.

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BREATH HOLDING IN INFANTS*

Breath holding (also known as hysterical laryngospasm, rage spasm, and inspiratory apnea) is a condition not infrequently observed in early childhood. At first glance one would be led to believe that the attacks are manifestations of spasmophilia or tetany with the associated laryngismus stridulus, but on closer investigation it will be discovered that such assumptions are erroneous. The breath holding attacks, as distinguished from laryngismus stridulus, manifest no true laryngeal spasm. The breathing stops suddenly in the midst of a crying attack, but there is no inspiratory spasm.

The attack may be briefly described as follows: The child stops breathing while he is crying in rage after prolonged expiration. He cries for a time, and then suddenly stops, finding it impossible, for a brief period, to make any further sound. The inspiratory muscles remain in a tonic state. The child throws himself about and becomes cyanotic or pale. Then the body becomes rigid and the eyes turn or become set. For a moment it seems that the child is asphyxiated. The attack usually lasts a few seconds and then he revives. It may, however, continue for several minutes, while in the more severe cases it may be followed by convulsions or unconsciousness.

Barthez and Rilliet were among the first to speak of breath holding. They describe it as a condition occurring in passionate (angered) children, manifested by a suspension of the respiration accompanied by suffocating inspiration, during which the face becomes purplish and the extremities cold. Sometimes it is followed by an eclamptic attack. They say it is difficult to distinguish breath holding from glottis spasm.

Ibrahim reports three cases, the first two of which are dismissed with a short reference. In the first there had been laryngismus stridulus of the ordinary type in early infancy. In both, the rage spasms were characterized by emotional excitation, cyanosis, unconsciousness, and great drowsiness. The third case is described in more detail, the principal points of which were the following: The mother was an extremely nervous woman and is said to have suffered from rage spasm during infancy. The infant thus had a nervous stigma, although there was neither epilepsy, insanity, nor other nervous disease in the family history. Ibrahim had this baby under observation since earliest infancy. During the second year, the child in the midst of a crying attack would hold its breath and become cyanotic; the body would grow limp. Respiration would apparently cease. At the end of the attack, the child would cry lustily for a few moments and soon afterward fall asleep.

Ibrahim calls attention to the fact that all the classical signs of tetany, including the electrical reactions, were searched for but were not found.

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* Insert by General Editor.

H. Neumann* in his discussion of breath holding, tries to differentiate it from tetany. Laryngismus stridulus, he says, is a manifestation of spasmophilic diathesis or tetany, and tends to disappear during the first year or two of life; whereas the inspiratory apnea or breath holding attack is an early symptom of neuropathic diathesis. These attacks especially during pre school age, though they may occur during infancy. He also says that in numerous cases which he has examined, the classical signs of tetany were usually absent. There was one exception, however, a boy of 4 years, who, suffering evidently from rage spasm, showed very definitely Chvostek's facial phenomenon. His father, who was an extremely neuropathic individual, showed the same sign.

A further differentiation may be made in addition to Neumann's observations. Attacks of breath holding occur commonly in the second and third years of life and tend to disappear about the fifth; whereas genuine spasmophilia makes its appearance during the first year or two.

It will occur to overyone that the resemblance between these breath holding attacks and minor epilepsy is such that the differentiation of the two may at times present great difficulties. It should be remembered, however, that breath holding attacks follow immediately after severe crying, excitement, or anger; while epileptic attacks occur suddenly in the midst of quiet play or even during sleep. Biting of the tongue, which is characteristic of epilepsy, does not occur in breath holding. On the other hand, involuntary evacuation of the bladder or rectum may occur in either attack. Another point of resemblance is the prolonged sleep usually following an attack, which is characteristic of both conditions.

Neumann reports a case in which breath holding attacks or rage spasm occurred in an epileptic child 3 years old. He had suffered from epilepsy since his third month of life, and presented, at times, unmistakable convulsive seizures. On certain occasions, however, when he lost his temper or became frightened, he would hold his breath, become cyanotic, and after a brief period would lapse into a deep sleep.

It would seem that an important point in the differentiation of the two conditions is to be noted in each case in the progress of the disease, namely, that breath holding attacks tend to recover completely, while epilepsy, as a rule, persists.

Attacks of whooping cough in young infants and children may require differentiation from breath holding seizures. A child was brought to the clinic because of peculiar respiratory attacks that alternated with coughing. At night the child nor infrequently had severe coughing attacks which the parents thought resembled pertussis. During the day, however, the coughing was substituted by paroxysms of breath holding, undoubtedly they were voluntary attempts to suppress the cough. They were characterized by extreme cyanosis, great prostration, relaxation of muscles, and momentary unconsciousness. The baby gives one or two short coughs, then becomes blue and rigid. She has a facial spasm, the eyes become set and roll upward, arms and legs are rigid, and the mouth is tightly closed. The attack lasts several minutes. Cyanosis disappears, normal breathing is resumed, and consciousness returns. This seventeen months old baby had such an attack in the clinic. These alternating attacks of coughing and breath holding had been going on for four or five weeks, during which time one or more typical whooping paroxysms occurred almost nightly.

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* Arch. f. Kinderh., 42.

A baby 2 weeks old acquired whooping cough from the mother. The child began to cough when it was only 10 days old. Each paroxysm was characterized by a slight crowing. He would become intensely cyanotic, and fall limp in the mother's arms with complete respiratory cessation. At such times, the pulse was slow and the child seemed lifeless. After a few moments the patient regained color, and breathing proceeded normally. Ultimately the baby recovered.

Cases of breath holding are not of uncommon occurrence. Very often they are the cause for emergency calls for the medical attendant, though as a rule, he discovers when he arrives on the scene that the child has recovered. A mother came to the clinic with a large, well developed baby of 22 months, who presented all the characteristics of breath holding or rage spasm. The mother described in minutest detail all of the points which have already been referred to. If the child was crossed or thwarted in any way, it would cry, hold its breath, become cyanosed and momentarily unconscious. The child was very well nourished, presented no evidence of rickets, no carpopedal spasm, and no Chvostek or Trousseau signs.

Another case further illustrates the disorder. A mother brought her 17 months old baby to us on May 5, 1918, because of anemia and great fretfulness. The child had always been sick and delicate, was very difficult to feed, and suffered from severe malnutrition. She was extremely irritable and cried much of the time. About two weeks before we saw her the child began to fret more than usual, would become extremely irritable and work herself into a rage, which would be interrupted by a breath holding spasm. The mother said that five or six of these attacks occurred daily. None of the typical signs of tetany was present, though there was a slight increase of electrical excitability. We improved the child's feeding, and suggested a plan of psychotherapeutic treatment which the mother, who was a very intelligent woman, immediately adopted. She refused to pick up the child when she cried, treated her sternly, shook her finger at her, and reproved her when a breath holding attack was imminent or in progress. The treatment finally proved successful. Although on the first day after the baby's admission to the hospital we were enabled to witness several typical attacks which consisted of fits of temper, crying, breath holding, cyanosis and momentary unconsciousness, on the second day they had completely ceased.

These attacks of breath holding occur in neuropathic children and are brought on by fear, anger, fright, or some other psychic trauma. As a rule, children subject to such seizures are irritable and ill-tempered. After the attacks have been once established, the child at times appears to utilize them to gain his end or wishes, later on they seem to occur involuntarily after fright or excitement or rage. The condition is very often aggravated by neuropathic parents who, fearing the recurrences of these attacks, humor them in every possible way, permitting no one to cross the child and allowing no whim or caprice to remain ungratified. It is obvious, of course, that such treatment does not help the child to build up any degree of resistance to the recurring attacks.

After some time has elapsed, most of these patients practically recover, spontaneously, although in some instances, as has been pointed out, they may be severe and protracted. Children who fall ill with an acute infectious disease, or who, by reason of accident or injury, require surgical treatment, may be seized with a severe breath holding attack, accompanied by general covulsions, as a result of the fright which they have sustained. Under these circumstances the attack may be so severe as to terminate fatally.

Treatment should be directed toward the general management of the nervous child. Mental stimulation of every kind should be avoided. He should be disregarded as much as possible by parents and friends. When the attacks occur, there should be no fussiness or consternation, or any of the hysterical manifestations usually evinced by mother or nurse. Cold water dashed into the face has been suggested as a direct remedy. Finally, the patient should be shown in no uncertain manner that any repetition of the attack will meet with harsh, if not painful, measures. Diverting the child's attention suddenly may prevent an attack.

Frequently the patients improve most rapidly when they are removed from the excitement and tension of home surroundings. A short stay at the hospital away from parents and friends is often advisable. At times a short course of bromide, or luminal treatment with calcium chloride of lactate may be required in addition to psychotherapy.

PICA*

Definition.—The ancients described this condition under the Greek name "kitta," signifying a bird of variegated colors which showed a marked propensity for eating all sorts of substances. "Pica" comes from the Latin word meaning magpie, a bird which shows a vitiated craving for what is unfit for food. Volpato described the endemic occurrence of this condition in Italy. He designated it "allotriophagy," a craving for unnatural substances.

The condition seems to occur more particularly in certain regions, for example, the clay eaters in the southern states of America. The

*Insert by General Editor.

dirt eaters of hook worm districts illustrate a perversion of appetite. These patients eat many gritty substances, particularly mud, clay, and chalk.

Children who suffer from this habit have an unnatural appetite for substances which are not intended for nutrition and are often repulsive or disgusting. Among the substances which are eaten are the following: chalk, plaster, sand, hair, wool, slate, graphite, uncooked rice, sealing wax, soap, tallow, paint from woodwork and furniture, gravel, coal, cinders, paper, and even fecal matter. This morbid appetite has been observed both in human beings and in certain animals, especially pigs and anemic lambs.

Parorexia.—It has been suggested that the term "parorexia" be used to characterize the various perversions of appetite. This condition may be subdivided according to the nature of the substances which are ingested: (1) "Pica" indicates a desire for articles which are not foods, such as chalk, clay, and earth; (2) "Malacia" is used to indicate a desire for spiced foods, vinegar, and mustard; (3) "Allotriophagy" has been used in designating a desire for disgusting substances, such as urine and fecal matter. The conditions designated as pica and malacia may exist in the same person, while malacia is a state of appetite found sometimes in chlorotic girls and sometimes in pregnant women. Allotriophagy is observed among idiots and insane people.

Age Incidence.—Doctor John Thomson made an interesting study, and found that the habit occurred frequently in children from 4 to 18 months or even later. Still reports that in nine of his fourteen cases the habit began in the second year of life. In one it began at eight months; in two it began in the fourth year.

Etiology.—There has been much speculation as to the cause of this condition. It has been thought that deterioration of the general health, particularly where marked anemia was present, was an active causal condition. Others have suggested that a cachectic class of children, or those suffering from rickets and protracted bronchitis, or from round worms, hook worms, malignant tumor, or tuberculosis, are likely to develop pica. Chronic gastro-intestinal catarrh has been suggested as a possible cause of this perversion of appetite. Nervous temperament or neuropathic constitution is considered the cause by numerous authors. Mentally deficient children sometimes show this peculiarity of appetite. Despite these various views as to the cause of pica, nothing is definitely known as to the exact etiology.

Symptomatology.—Whatever is the cause of this perversion, we know that after a time the patient acquires a habit which leads him to eat more and more of gritty substances, and the quantity continues to increase, in order that the perverted appetite may be satisfied. Young infants who suffer from this habit have milder manifestations than those which occur in later childhood or adult life. Many of these patients suffer from some form of gastro-intestinal disturbance and anemia.

One of the most striking symptoms consists of the tendency to diminish the normal appetite, and to increase the quantity of dirt eaten. When the food is materially diminished, and the amount of dirt which is eaten assumes a considerable proportion, it is to be observed that the children become anemic, malnourished, and nervous. Constipation is not infrequent. Sometimes it may become so severe as to suggest a mechanical obstruction of the bowels. Observation, however, seems to show that the nutritional disturbances of pica are more marked in older children and in adults than in the young infant. It has been noted that the negroes of Jamaica sometimes carry this habit to great excess. If this practice is long continued, they succumb.

In the infantile type of pica, it is found that the condition is sometimes associated with gastro-intestinal disease and worms. A case is reported by Bohn, in which is described a child of two years who bit off and chewed her own blond hair. In each stool this matted hair could be seen. This patient lost very much in weight and finally died of inanition. Benson* reported a case of a girl of seven years, who suddenly became ill with headache and nausea, followed by vomiting and frequent bloody stools. Three days later she passed the tip of an ordinary kitchen knife. The diarrhea and bloody stools persisted for several weeks, but the child made an uneventful recovery.

It has already been noted that pica may occur in early infancy. It has been observed that young babies, before they are able to walk, will lick the mud from the fathers' boots, or the dirt and gravel from the wheels of their own ambulators. Later on, they pull the plaster off the walls, and subsequently begin to eat all sorts of things. Many of these children are thin, pale and delicate in appearance. Another group has colic and diarrhea, and the stools frequently contain clay, sand, and other substances which have been ingested.

I recall two cases in which children ate the paint from the household furniture. One of them presented symptoms of lead intoxication. Neuritis and lead encephalitis may occur. Recently a child of two years, who eats almost any substance that she can lay her hands upon, came under notice. The latest episode consisted of eating glass beads and face rouge. Some infants choose only one unsuitable substance, while others partake of several substances. Still others select a large menu of undigestible and gritty things of every conceivable kind.

Prognosis.—In infancy the prognosis is usually good. This is also true in children under four years of age. It has been suggested that the average duration of these cases is estimated at twenty months. The early infantile type tends to spontaneous recovery during the third or fourth year, especially when the intelligence of the child awakens.

Cases are cited, however, in which the habit persists for a long period, often continuing until adolescence. As a general rule, no serious harm follows, though fatal cases have been reported from eating sand, gravel, or hair. Diarrhea sometimes occurs, though constipation and fecal impaction are not infrequently observed.

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* New York State Medical Journal, 1919.

Treatment.—Thomson states the indications for treatment as follows:

1. Keep the child away from the substances for which he has a morbid craving. All habits are strengthened by practice, and their hold slackens under disuse.
2. Treat the digestion. Any local or general uneasiness tends to increase the craving.
3. Improve the general health. These habits have a far stronger hold on the weakly; the strong throw then off readily.
4. If possible, change the child's surroundings and occupy his mind with new interests. Let him be kept happy and busy.

LEFT-HANDEDNESS

Left-handedness is a natural state in which a person possessing a perfectly normal right hand prefers to use his left. Some right-handed persons acquire an ability to use the left hand with remarkable dexterity. Under circumstances in which the right hand is rendered useless, as from paralysis or other lesion, the left hand is called upon to assume the functions of the right. Such cases cannot be classed as left-handedness because the natural tendency is to use the right hand, as becomes manifest when especial skill is required in the performance of an act. Special acts, such as saluting, bread cutting or ball throwing may be used as a test in estimating the approximate percentage of left-handed persons. The percentage of left-handedness is higher in small children than in adults, because children have not had sufficient opportunity to learn to mask their left-handedness by education, imitation, and drilling. In adults the percentage is about 5 per cent. (Ogle,[1] Stier,[2] Bardeleben[3]), and there is a smaller group of so-called ambidextrous persons who use both hands with equal facility. Males are more frequently left-handed than females. Of 17,074 children examined by Schaeffer[4], 4.06 per cent. were left-handed (boys, 5.15 per cent.; girls, 2.98 per cent.). Bardeleben estimates that 9.5 per cent. and Stier that 7.4 per cent. of cases occur in males. Schaeffer estimates that boys constitute 62.6 per cent. and girls 37.4 per cent. of left-handed persons in general.

A left-handed person seems to possess greater skill in his left leg as well. It was also proved by anthropologic, psychologic, and clinical investigation that even vision, hearing, the general sensibility, the weight, size, and volume of the bones, the muscles, blood pressure, mimical expressions, etc., are better developed on the left than on the right side in left-handed persons, the reverse obtaining in a right-handed person (Biervliet).[5] Ogle asserts that in these cases the right half of the brain, controlling the left side of the body, is structurally better developed than the other half, as regards weight, amount of convolutions, gray matter, etc.; in the right-handed, the opposite obtains. Though many of the foregoing statements are hypothetical and not scientifically proved, the fact remains that a left-handed person possesses a physiologically superior left side, a peculiarity of very ancient origin, that was evidently common among prehistoric races. Thus de Mortillet[6] demonstrated by prehistoric implements found in France, that left-handed persons were twice as numerous there as right-handed persons, while Ogle observed left-handedness even in animals (apes) and bird (parrots). The majority of authorities consider left-handedness as belonging to the human race only, denying its possible occurrence in animals.

Etiology.—The causes of left-handedness and right-handedness have occupied the minds of anatomists, anthropologists, psychologists, neurologists and philosophers for many centuries. Numerous theories, or rather hypotheses, have been advanced, such as heredity, influence of environment, habits, imitation, natural laws of evolution, a richer blood supply of one hemisphere, and transposition of the viscera. For instance, association of a child with a left-handed nurse is known to have resulted in the child's becoming left-handed. After a right-handed nurse was employed, the child became right-handed.

The habit of carrying the child on the left side is supposed to give its left hand better chances and greater freedom to develop. In this connection, Feltz's[7] case is unique. Two left-handed children, without faulty heredity, became right-handed when the habit of carrying them on the left side was changed. Rothschild's[8] experience shows that imitation may be the cause, since, in his case, through suggestion, the left-handedness disappeared permanently. The most plausible cause is heredity (in 50 per cent. of cases according to Stier), direct or indirect, near or remote, as this condition could be traced from family to family, and among the collateral branches. For instance, Cunningham[9] quotes a case in which five children were born of a left-handed father and a left-handed mother, four of whom were left-handed, and one, a girl, was right-handed; three cousins of the family were left-handed. In another case, in which the mother was left-handed and the father was right-handed, all of the 14 children (eight brothers and six sisters) were left-handed. The mother's father, two sisters, and three brothers were left-handed. The father's brother was left-handed and he had five left-handed children.

Some authors (Pye Smith,[10] Stier, Weber) look upon right-handedness and left-handedness as remnants of a primitive, dying-out peculiarity. According to this hypothesis, the primitive human creatures, in their fights with beasts or with one another, used either left or right hand. Those that used the right hand were more successful, as they protected the heart, with the other hand, and thus encountered fewer dangers and avoided mortal wounds of the heart. The opponents who used the left hand in fight were doomed, as they could not protect the heart, neither were they able to inflict mortal blows in that region on their foes. The users of the right hand, therefore, had a better chance to survive in the struggle for existence, the greater majority of the human race being descendants from these successful right-handed fighters, the minority, the left-handers, being descendants of the left-handed fighters. Whether the primitive fighters were led by instinct, or by knowledge of anatomy and physiology, to protect their heart with the left hand and fight with the right is, of course, not known.

Hartshorne,[11] Gould, Stevens and Ducasse[12] offer another hypothesis. In their opinion, on account of a more powerful development of the left half of the retina, images coming from the right field of vision are brighter and larger, thus attracting the attention of the child. It grasps eagerly, therefore, with the right hand, everything coming from the right, which by earliest experience acquires a special skill. According to this theory, the opposite, of course, obtains in left-handed children. It is needless to enter into detailed discussions of the numerous theories and hypotheses, as none really explains the cause of right-handedness or left-handedness. For instance, Stevens and Ducasse merely state the hypothesis that the left half of the retina is better developed. But this better development is the result of the preëminence or superiority of the left hemisphere in general, the superiority being responsible for the better development of the right side of the body, including speech, ability to write, etc. Some authorities claim that even such a view is wrong, that in the newborn both hemispheres are functionally alike, and that the right hand becomes better developed because it has been exercised for ages, while the left hand became neglected, which fact resulted in breeding right-handed persons. According to this teaching, the right-handed person cannot be considered normal either, for his left side is functionally inferior. A normal condition exists only when both sides are equally well developed, as in ambidexterity.

Notwithstanding such able and plausible arguments, one must agree with Biervliet that the real cause of right-handedness or left-handedness will long remain unknown, and that if the cause is related to the embryonic deficiencies of the human body, it will never be found.

Pathophysiology.—As noted, the left side of the body in a left-handed person is better developed physiologically and anatomically than the opposite side, owing to the superior development of the right half of the brain. In left-handedness the functions usually belonging to the left side of the brain are transferred to the right, the left-handed person functioning principally through the right hemisphere. Apoplectic strokes, from hemorrhages, thrombosis, etc., producing hemiplegia on the left will be accompanied in a left-handed person by aphasia, agraphia, etc. This is not true in right-handedness.

It should be mentioned also that stammering, stuttering, and, mirror writing are frequently observed in left-handed children. Schaefer found 2.3 per cent. stutterers among 692 left-handed children. This he thinks is because stutterers have their speech center developed in the left, while it should be better developed in the right hemisphere.

The **diagnosis** is based on history—whether the peculiarity dates from early childhood, whether there is a family tendency (direct or indirect) to use the left hand, and whether it can be noticed at eating or in ball throwing, ball tossing, threading a needle, and similar more or less skilful acts. By means of the latter, it is possible to diagnose left-handedness even in cases masked by education, imitation, proper drilling, etc.

The **prognosis** depends largely upon the severity of the anomaly, there being extreme, moderate, and mild cases. The latter two forms, by process of education, may be transformed into right-handedness, or ambidexterity, while the severe, obstinate form defies any methods of training.

Treatment.—A pronounced left-hander should not be forced to use his right hand. On the contrary, he should be encouraged to use the left one, especially in writing, for this seems to have a great influence on the development of his right hemisphere, as it is kept in better and more continuous development by persistent use of the left organs. In other words, the functions of the better half of the brain are thus constantly promoted, while in using the right hand, a left-handed person would make demands upon a portion of the brain which, being naturally inferior, cannot give, so to speak, such good service as the properly developed right one. Ireland[13] justly remarks that a left-handed child, forced to write with his right hand through fear of punishment, is very much in the same position as a right-handed one who should be forced to hold his pen in the left. Bardeleben says that attempts to convert a left-handed person into a right-handed person are, in left-handers, the causes of speech troubles, such as stuttering. It is, therefore, imprudent to attempt to transfer a speech center from one-half of the brain to the other; in fact this is impossible.

In spite of soundness of such arguments, a large number[14, 15, 16, 17] advocate ambidexterity. In their opinion, both hands should possess the same physiologic skill; in other words, every person, in their opinion, should be ambidextrous, for an ambidextrous person, they say, must necessarily be more accomplished, better fit for life's struggles, and of greater use to the community.

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HEMIHYPERTROPHY

Hemihypertrophy, literally, means unilateral enlargement of the body, including the skull, face, abdomen, upper and lower limbs, the nose, jaws, palate, and even the large vessels (carotids in Cagiati's case[1]). In a minor group of cases the hypertrophy is confined to but one of the foregoing parts (partial hemihypertrophy), while in a third group of cases involving segments on both sides, the hypertrophy is crossed. In Lewin's[2] case, for example, hypertrophy of the face and arm existed on the left side, and hypertrophy of the leg on the right. In the case of André Thomas,[3] hypertrophy of the right side of the face and tongue was associated with that of the trunk and leg on the left side. Whatever the variety, the hypertrophy involves all of the tissues included in the affected part of the body, such as the skin, subcutaneous tissue, bones, muscles, and even the nerves. The contrast between the *enlarged* segment and the corresponding *normal* one is so striking, especially in typical cases, that the body seems to be made up of two parts, belonging to two different persons.

In less typical cases, the discrepancy is not so pronounced, and measurements are necessary to establish the differences, which otherwise might be overlooked.

Changes in the affected segment are both anatomic and physiologic, resulting in an alteration in length, size and power. The skin is soft, does not pit on pressure, perspires normally or profusely, and frequently exhibits vascular and trophic changes. To the former belong increased arterial pulsation (Bassoe[4]), hyperemia, nevi, varices, marbling; to the latter, abnormal hair growth, lanugo hair on the nose and ear, and increased sebaceous and sweat-secretion. The temperature on the affected side may be slightly increased, while the sensibility, the galvanofaradic conductibility, may be somewhat diminished. Subjective complaints or motor disturbances are absent, except in cases in which an unusually hypertrophied foot or toe interferes with function. As a rule, however, in spite of the difference in the size, walking, jumping, dancing, and running are possible.

In many cases, as pointed out by Trélat and Monod,[5] limping is not manifest, the victim compensating for the discrepancy in the size of the legs by keeping the overgrown limb slightly bent, and the pelvis on the corresponding side somewhat elevated.

The various types of hypertrophy have been observed more in men than in women, more frequently on the right than on the left side, and either immediately after birth (Redard,[6] Kiwull,[7] Davidson[8]) or much later in life. Some authors (Trélat and Monod and Fischer[9]) consider this anomaly as congenital, Mackay[10] believes that the condition is acquired, while Sabrazés and Cabannes[11] admit both possibilities. Whether congenital or acquired, the hemihypertrophy always progresses, one-half of the body steadily growing faster than the other side, causing no real suffering, and even little deformity, except when the face or the toes are much affected. Facial hemihypertrophy, is of particular importance, since it possesses features

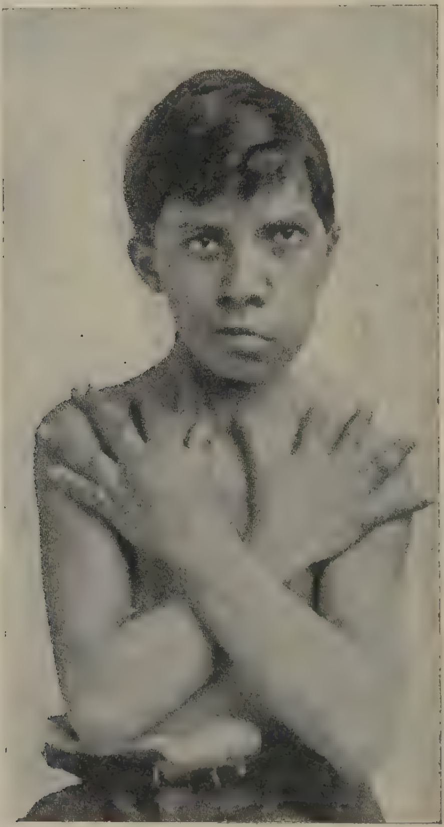

Fig. 40.—Hypertrophy of the left side of the face, especially of the cheek. The larger size of the left forearm, hand and fingers is also marked. (From Bassoe, Peter: Am. J. Insan.)

the reverse of those in facial hemiatrophy (Fig. 40). In facial hemihypertrophy the affected cheek is enlarged, appearing to be puffed up and elevated; the point of the nose is deviated to the opposite side; the nasolabial fold is raised; the alæ nasi are drawn toward the deformed side; one-half of the lip is thicker; the teeth are more fully developed than on the normal side. For instance, in Finlayson's[12] case, there was a total right hypertrophy, with involvement of the right half of the face, arm, leg, and toes. The patient had eight teeth on the right side and none on the left; the first tooth was out when the patient was three months old, and at the age of eighteen months, there were five teeth in the right upper jaw and three in the right lower jaw.

The hypertrophy may involve one-half of the tongue, the normal portion appearing as a mere appendage to the hypertrophied portion; or the ear on the affected side may be much larger than on the side that is not affected. The resulting deformity of the face causes a marked contrast in the facial expression of the affected and non-affected sides.

Histologic studies of tissues excised during the patient's life (Kiwull, Montgomery[13]), as well as post-mortem examination (Busch,[14] Hornstein,[15] Arnheim,[16] Cagiati[1]), revealed that the hypertrophy is caused by hyperplasia of the connective and adipose tissues, and of the sweat and sebaceous glands. The parenchyma of the nerve tissue was found intact but surrounded by a thickened and proliferated connective tissue (endoneurium, perineurium and epineurium). Arnheim and Cagiati also directed attention to the increased size of the vessels, hyperplasia of the muscles, liver, and kidneys. No change has been described as occurring in the brain and the sympathetic nervous system.

The diagnosis of hemihypertrophy is simple. A comparative measurement of both sides will reveal an abnormal length of either an entire limb or its segment. The malleoli or the knees are not on the same level; the circumference of one limb is larger, or the face may be deformed. The presence of vascular changes, such as nevi and varices, assists in the diagnosis. Hemihypertrophy must be differentiated from hemiatrophy, chronic trophedema of Meige, elephantiasis, acromegaly, and syringomyelia. The last three diseases are rare in children. The healthy side appears smaller and the condition may simulate atrophy, which should be excluded by the presence of normal muscular power, normal nerve responses, and the absence of flabbiness and disturbed function. Chronic trophedema of Meige is a hereditary or family disease without bone involvement, and the edema which characterizes trophedema is absent in unilateral hypertrophy.

The causes of this anomaly are wholly unknown, but it seems certain that syphilis, heredity, and obstetric anomalies play no etiologic rôle. In some cases a history of trifacial neuralgia, otitis with a concomitant infection, or trauma of the mother during pregnancy has been obtained. The association is certainly accidental, and fails to explain why only one portion of the body should, during the whole of the patient's life, continue constantly to grow faster than the other side. Trélat and Monod explained the abnormal growth by assuming a partial paralysis of the vasomotor or sympathetic nervous system, with resulting venous stasis in the affected parts. Hermanides,[17] Ziehen,[18] and others, attempted an explanation on the basis of an abnormality of the Gasserian ganglion, while Lewin, Mackay and Piazza[19] promulgated the so-called trophic theory, according to which the trouble is supposed to lie in the trophic centers and nerves controlling growth. As the laws of growth are wholly unknown, the foregoing theories are untenable. It should, however, be pointed out that there are many other morbid conditions in which the laws of growth are at fault. Here belong gigantism, dwarfism, acromegaly, and various forms of dispituitarism, all of which are associated with definite disturbances in the pituitary and pineal glands. It would be proper to include hemihypertrophy in the foregoing groups. Dana's[20] case of acromegaly associated with hemihypertrophy, the cases of Chassaignac,[21] Fischer, Friedberg,[22] Hornstein, and others, in which the hemihypertrophy was so great that it closely resembled gigantism, possibly show the identity of these morbid conditions. In Friedberg's case, for instance, the increased size affected, as in acromegaly, the terminal portions of the lower extremity, decreasing toward the inguinal region. It is possible that some types of hemihypertrophy may be manifestations of acromegaly, in a modified, incomplete, or rudimentary form.

The prognosis for life is good, patients usually succumbing to some intercurrent disease. In one case (that of André Thomas), there was observed within 18 months a decrease in the volume of some portions of the body, while some parts attained their normal size.

Treatment is indicated only when there is manifest discomfort, which, in the case of a toe or finger, for instance, may necessitate amputation (Fischer,[9] Prince[23]). In other cases a high shoe for the healthy foot is of great benefit to the patient in walking. Redard[6] recommends gradual compression of the hypertrophied organ by elastic bands. Pituitary extract may be tried, but at this time reports are insufficient to judge of its value.

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HEMIATROPHY (ROMBERG'S DISEASE)

Hemiatrophy, or unilateral wasting, rarely affects the entire half of the body. Since the condition is usually partial, and involves, as a rule, only one side of the face, it is commonly known as facial hemiatrophy. On account of its progressive course, Eulenburg[1] called it progressive facial hemiatrophy, a term used by the majority of authors. The anomaly usually becomes manifest before the age of 10. Bechterew's[2] patient was four years of age, which is the earliest age so far recorded. Girls are affected more frequently than boys, and the left side is more commonly affected than the right (Stier[3]).

The atrophy usually begins on the cheek or some other part of the face (forehead, nose, eyelids), as a whitish discoloration. Several such islands of tissue may coalesce and, by a gradual increase of size, form larger areas deprived of pigment, but capable of blushing from emotion or artificial stimulation (Eulenburg).

In advanced cases, the affected skin becomes thin, emaciated, deprived of panniculus adiposus, and closely covers the subjacent bones (malar, temporal, upper and lower jaws). The latter may participate in the atrophy, depending, as Byrom Bramwell[4] suggested, on the time of the occurrence of the atrophy. If the condition begins early in life, before the bone growth is an accomplished process, then atrophy of the bones of the face will obtain. In case the bone skeleton is already developed, the atrophy of the facial bones will either be slight or altogether lacking. The atrophy also involves the sweat and lacrimal glands, the parotids, the nasal cartilages, and the ear, while the muscles principally supplied by the fifth nerve, may remain unaffected.

The atrophy results in a facial asymmetry which, when examined from the side, gives to the face the appearance of consisting of two different halves. On the atrophied half, the cheek is concave, the nostril appearing smaller with its cartilage turned toward the affected side, the mouth somewhat open at the corner, the nasolabial fold sharper. The hair on the affected side becomes brittle, coarse and discolored. It may turn white, with arrest of growth, and not infrequently it falls out. The teeth occasionally become loose, and finally also fall out. Sensory and electrical changes, trophic disorders, such as ulcers and blisters, subjective complaints of pain, paresthesias, disturbances of smell, hearing, and taste are very unusual. In many cases, the atrophy affects also the opposite side of the face (Fischer,[5] Oppenheim,[6] Wolff[7]), and is confined, as in the classical hemiatrophy, to the distribution of the fifth nerve. The process may go beyond the distribution of the fifth nerve and invade the regions of the seventh, twelfth, and even the tenth nerves. In Hutchinson's[8] case, for example, half of the soft palate was atrophied. The clinical picture, in such cases, will be complicated by atrophy of the lips on one side, and of one-half of the tongue, with deviation of the tongue toward the diseased side. The atrophy may spread to other parts of the body, involving the arm, leg and trunk, causing a total hemiatrophy, as described by Raymond and Sicard,[9] Orbison,[10] Hutchinson, Burrel,[11] and others. In still rarer cases, the atrophy does not begin in the face, but has its origin elsewhere in the body, invading the face afterward (Raymond-Sicard); or the origin is facial and later involves the opposite arm and leg (*crossed* hemiatrophy, Luntz[12]).

The differential diagnosis is to be made from congenital muscular deficiency, arrest of muscle growth caused by injuries, progressive muscular dystrophy (type Duchenne-Dejerine-Landouzy), tabes, and syringomyelia. The unilateral involvement, combined with the change in the color of the skin, the peculiarities in the growth of hair, and other features establish the diagnosis.

Histopathologic changes are confined to atrophy of the connective tissue of the skin, partly of the bones, and sometimes of the muscles (Hammond[13]). Mendel[14] found an interstitial neuritis of the fifth and musculospiral nerves in his case, with degeneration of the spinal root of the fifth and of the substantia ferruginea. Clinically, there was involvement of the areas supplied by the three branches of the trifacial nerve, the scapular region, and the forearm. Neither Mendel's findings, nor pathologic studies by other observers (Loebl and Wiesel[15]) explain the nature of this disease. The theories in vogue are mostly physiologic, namely the trophic theory of Romberg, Hitzig,[16] and others, and the angioneurotic or sympathetic theory as set forth by Bergson, Oppenheim, and Bouveyron.[17] Trophic centers and nerves are supposed to be at fault according to the former theory, while disturbances of the sympathetic nervous system are assumed to be causal in the latter. Claude and Sézary[18] found a pleocytosis in the spinal fluid and maintained that the process was of meningeal origin. It is certain that, in hemiatrophy, trophic disturbances dominate the clinical picture in a manner very much resembling scleroderma.

Hutchinson, who was one of the earlier observers, defined this syndrome as "nothing more than the arrest of growth following an attack of fifth nerve morphea, which occurred early in life." Cassirer[19] also looks upon hemiatrophy as a scleroderma confined to the area of the fifth nerve. In other words, this anomaly may be classified as a *trophoneurosis*.

The cause of hemiatrophy is unknown. Injuries and infectious diseases are usually mentioned as etiologic factors, while syphilis and various intoxications have no direct association with the disease. Heredity is rarely a factor in etiology, although it was apparent in Klingman's[20] case, in which physical deviations occurred in successive generations—the grandmother, mother, and two daughters, who were twins. One of the daughters developed a complete unilateral atrophy of the right side of the face; the other had a noticeable asymmetry of the face and ears. Klingman and Bannister[21] suggest frostbite and epilepsy as causative factors, while Möbius[22] wrote of the association between hemiatrophy and infection of the tonsils, or foci of infection in the skin and mucous membranes. Many cases have occurred following scarlet fever, erysipelas of the face, tonsillitis, and infected teeth.

In regard to prognosis it may be said that there is no danger to health or life from this disease. The course is protracted; but sooner or later the process is arrested. Some authors (Bramwell, Tanturri, Sèzary and Claude) report improvement from treatment.

In view of the possible infectious origin of this disease, prophylactic treatment of tonsils and decayed teeth, as first advocated by Möbius, is strongly recommended. When once the disease is established the treatment is symptomatic: Massage, electricity, galvanization and paraffin injections,[23] or injections of thiosinamin 0.02. c.c. every two days for 12 injections are recommended. Some clinicians prescribe potassium iodide while others, among them Dercum,[24] resort to resection of the branches of the fifth nerve.

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JUVENILE PARESIS (JUVENILE PARALYTIC DEMENTIA; PROGRESSIVE PARALYSIS OF THE INSANE IN CHILDREN)

Progressive paralysis of the insane occurs rarely in children, although its occurrence is not so infrequent as that of juvenile tabes. During six years, Alzheimer[1] observed but three cases of juvenile paresis among 360 adults, and Thiry[2] saw three cases among 7000 children, in four years, while older observers, such as Mendel and Christian, doubted its occurrence in children altogether. Since Clouston's[3] first record of a case of juvenile paresis in 1872, numerous other reports have appeared, the number at present running into the hundreds. The general conclusion is that etiologically, clinically, and pathologically, juvenile paresis in no way differs from the same malady in adults, though there may be some slight differences, as will be pointed out later.

Age.—Zappert[4] and Hunt[5] report that the earliest age of definite onset was six years. Watson[6] gives the average age as 14, the time of puberty.

Sex.—It occurs with equal frequency in both sexes, though Rondoni[7] asserts that the number of cases among girls slightly predominates, while Arsimole and Halberstadt[8] claim that boys are more frequently affected (out of 205 cases, 118 were boys, 87 girls). In adults, men are invariably affected more often.

Symptoms.—As in the adult, these are both mental and physical. The former appear quite early. An apparently healthy and normal child begins to show signs of mental deficiency, such as loss of memory for past and recent events, lack of attention and interest in the surroundings, and lack of ability to learn. The victim does not play with other children, is apathetic, listless, indifferent, idle, or unusually excitable and morose. In some cases, there occurs spasmodic crying or laughing, similar to that of pseudobulbar paralysis. In short, diminished intelligence, especially memory defects, is an early, and sometimes the only, sign of juvenile paresis. Alzheimer says that this may be noticed at a very early age, long before other symptoms become more or less pronounced. In the course of the disease, other mental symptoms, such as euphoria, hypochondriasis, stuporous or depressed states, delusions and hallucinations, may be observed, but much less frequently than in adults. According to Babonneix,[9] these symptoms occur in about 12 per cent. of cases, the delusions of grandeur being of a childish, puerile nature, like the slightly exaggerated ideas of a normal child. They pertain, for instance, to toys, eatables, etc. In two cases observed by Watson,[6] the delusions were of the adult grandiose type, these patients having been unusually exalted.

The *physical* symptoms, as a rule develop, simultaneously with the psychical. They are manifested first in retarded physical development. The patient is undersized and juvenile in appearance. Boys of 15 years or older may look like little children. The juvenile paretic otherwise shows physical anomalies similar to those of the adult. These are revealed in the speech, hand-writing, pupillary changes, the condition of the cerebral nerves, the muscles, reflexes, gait, and the general condition.

The *speech* becomes slurring, hesitating, and tremulous, with lapse of syllables. This is usually accompanied by twitchings in the facial muscles. It is indistinct and crippled, or dysarthric, finally becoming totally unintelligible, or anarthric. In the last stages, there is a total inability to speak, to utter a single articulate sound, or to understand words. In some cases the speech is slow, monotonous, and bradylalic, resembling somewhat the speech in multiple sclerosis; or the speech may be explosive.

The handwriting is greatly impaired and tremulous; the figures or letters are irregular, and letters are repeated. In the latest stages, writing is impossible, forgotten—there is complete agraphia.

Pupillary changes are important in the clinical picture of general paresis. The pupils may, however, in somec ases, be perfectly normal in size and in their reaction to light. In many instances, they are regular in shape, but unequal, one being larger than the other, the reaction to light being sluggish or lost. In other cases, the pupils are irregular in shape, misformed, and react poorly to light. In the majority of cases, they react in accommodation, but not to light (Argyll Robertson pupil). In still other instances, the pupils fail to react to light, accommodation and convergence. This condition is called iridoplegia. Any of the foregoing pupillary changes, when associated with mental disorders, are strongly suggestive of a brain lesion, especially that of general paresis.

The voluntary muscles may remain normal in nutrition and function, but they also may show signs of early involvement. In this condition tremor occurs, especially of the muscles of the face and tongue. The facial twitchings show most prominently during talking or any voluntary movement of the face. The fibrillary twitchings of the tongue may be seen clearly when it is protruded, or when the mouth is opened. In the further course of the disease, the general musculature of the body becomes atrophied; contractures occur which render standing, walking, or handling things impossible. In short, the patient becomes entirely helpless. In the advanced stage, involuntary muscular contractions are sometimes observed (flexion, extension or abduction of the fingers, rhythmic movements of the limbs, etc.).

The cerebral nerves are often involved, especially the second and third. The former is sometimes atrophied (optic atrophy), and involvement of the latter results in internal or external ophthalmoplegia (ptosis, strabismus). The reflexes show inconstant changes. As a rule, they are exaggerated and accompanied by clonuses or Babinski's sign, in some cases they remain normal, or are diminished. The gait is much impaired, frequently resembling that of cerebellar ataxia, the walk being unsteady, vacillating, and uncertain, the feet being held far apart. The general condition is usually affected to a great extent. There are severe headaches, and apoplectiform and epileptiform, seizures, which may be the earliest manifestation of juvenile paresis. In the latest stages, the general condition suffers still more. Atrophy of the muscles and subcutaneous fatty tissue becomes very marked. There are great debility, utter helplessness, and untidiness, which, in combination with the dementia, create a pitiful impression. Sensory disturbances may be present, but the mental state of the juvenile paretic is usually such that these cannot be properly determined by sensibility tests.

The clinical symptoms—mental and somatic—are sufficiently definite, at least in the majority of cases, to permit of a correct diagnosis. In doubtful cases, it can be confirmed by serologic examinations. In juvenile paresis, as in that of the adult, the blood gives a positive Wassermann reaction, while the spinal fluid shows, in adition, a pleocytosis (increased amount of cells), a positive globulin and positive Lange tests. Some authors (Hunt, for instance) lay especial stress on the Lange test, asserting that it is invariably positive in every case of general paresis.

Differential Diagnosis.—This should be made from (1) various forms of idiocy, (2) brain syphilis, (3) multiple sclerosis and (4) dementia precox.

1. Idiocy.—It was long ago suspected that many so-called idiots are, instead, sufferers from general paresis, and since the diagnosis of the latter disease is facilitated by the serologic tests, it is now possible to differentiate such idiocy from other forms, especially if a reliable history is obtainable and a careful physical examination reveals pupillary changes, and other characteristic findings.

2. Brain Syphilis.—General paresis may be combined with brain syphilis. The third nerve involvement shown in ptosis, for instance, may result from cerebral syphilis (meningitis), but as a rule, the combination of physical and mental symptoms is so characteristic, that a differential diagnosis between general paresis and various types of brain syphilis is rarely necessary.

3. Multiple Sclerosis.—In multiple sclerosis, there are signs similar to those of general paresis, and in adults the differential diagnosis may be exceedingly difficult. In children, since multiple sclerosis is a rare malady, it is easily eliminated, especially in the presence of the classical clinical picture and the serologic findings.

4. Dementia Precox.—This should be borne in mind in some cases, especially in the early stages, when the condition of the memory, emotional states, etc., resembles that of both these diseases. Here again, the clinical picture gives the key, especially pupillary changes. Serologic findings help in the diagnosis.

Etiology.—Syphilis, either hereditary or acquired, is the sole cause of this disease. The greatest majority of juvenile paretics show signs of hereditary syphilis, such as Hutchinson's teeth, chorioretinitis, keratitis, linear scars at the angle of the mouth, misshapen nose, etc. Very few cases of juvenile paresis are acquired from sexual intercourse, kissing, or other causes (Westphal's patient[10] was 15 years old and had acquired syphilis from intercourse at the age of eight). Regis[11] states that it may occur even as a purely familial disease, as when several children with hereditary syphilis ultimately become paretics. In some cases the parents themselves suffer from general paresis, as in MacDowall's cases.[12] In one case, four members of a family (brothers and sisters) were affected with general paresis at the ages of 20, 12, 18, and 20, respectively. In another case, general paresis occurred in the father, mother and son. Some authors see in such cases an abnormal predisposition or susceptibility of the nervous system, caused by the syphilitic virus. It has been proven that syphilis actually produced developmental anomalies in the nervous system, especially the cerebellum, which ultimately result in general paresis. The time between the syphilitic infection and the first manifestation of the disease is from 14 to 15 years, as in adults, which fact also confirms a syphilitic etiology.

Pathologic Anatomy.—The pathologic changes in juvenile paresis are of the same character as those in adults. In addition, in almost every case, developmental anomalies are found, notably in the cerebellum. The weight of the brain is decreased; the gray matter is atrophied; the pia is greatly thickened and adherent to the dura, as well as to the brain tissue. The convolutions are diminished in size; the sulci gap; the white fibers are greatly decreased in amount, and in some places totally absent. The subdural space and the ventricles are dilated and filled with fluid. The ependyma is granular; the ganglion cells, especially of the deeper cortical layers, are totally changed or destroyed; the glia is proliferated. The vessels are congested; their perivascular or adventitial spaces are infiltrated with plasma cells and lymphocytes. In addition, there is marked proliferation and formation of new capillaries. Besides plasma cells and lymphocytes, mast cells, fibroblasts, rod cells, and gitter cells are often found. Especially noteworty are the changes in the cerebellum, in which the usual mesodermic and ectodermic phenomena are combined with marked developmental anomalies. Thus, the Purkinje cells often show two or three nuclei; they are greatly increased in size, vacuolated or elongated, or two Purkinje cells may become welded. In some cases the Purkinje cells may be absent. According to Sträussler,[13] Lafora,[15] Trapet,[14] and Rondoni,[7] some are pigmented or sclerosed and some are misplaced toward the molecular layer of the cerebellum. Rondoni[7] and Vogt[16] describe ganglion cell proliferation in the cerebral cortex. The spinal cord shows the same changes as those found in the brain—thickened hyperplastic and infiltrated pia arachnoid, but the parenchymatous lesions take the form of degeneration of the pyramidal tracts.* Spirochetes found in the brains of adult paretics have not, so far as I am aware, been demonstrated in juvenile paresis.

Course and Prognosis.—The course is always chronic and progressive, lasting from one and one-half to nine years. The average duration is five years. In general, the course is more protracted and chronic in children than in adults, always ending in complete dementia. The mental and physical symptoms become progressively worse; the patient is confined to bed, is helpless, and has to be fed. Remissions are unusual; the course is uninterrupted and invariably ends in death. The prognosis is, therefore, absolutely hopeless.

Differential points[17] of diagnosis between juvenile paresis and general paralysis in adults are: (1) In juvenile paresis syphilis is congenital, seldom acquired; in adults, the opposite obtains. (2) Most of the patients are feebleminded before the disease fully develops; in adults this is not true. (3) The prodromal stage in juvenile paresis is unusually rich in symptoms, especially in epileptic attacks; in adults it is less so. (4) Delusions are rare; remissions are unknown in the juvenile form. (5) The duration is longer.

Treatment.—Prophylaxis is the only satisfactory treatment. A child with hereditary syphilis or born of syphilitic parents should be energetically treated in early infancy with mercury and arsphenamin. Even then, a complete cure cannot be expected in view of the developmental anomalies which obtain in hereditary syphilis. When the symptoms are fully developed commitment to an institution is necessary.

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AMAUROTIC FAMILY IDIOCIES

Since Bernard Sachs[1] of New York described, in 1887, a syndrome of blind idiocy bearing the name of Tay-Sachs' disease, several other forms have been described, differing, etiologically, clinically, and pathologically from the Tay-Sachs classical type. Amaurotic family idiocies may be properly spoken of and there are a number of clinical syndromes which are by no means identical, though they much resemble one another.

Tay=Sachs' or Infantile Type.—Clinically this type is characterized by a rapidly developing mental deterioration, blindness, and specific changes in the eye fundi. Pathologically, according to Schaffer definite changes appear in the ganglion cells of the entire central nervous system.

Symptoms.—The disease begins during the first six months of the child's life, usually between the ages of three and six months, seldom later. The first noticeable sign is listlessness or inactivity, the infant being indifferent to its surroundings. It does not seem to care for anything, does not attempt voluntary movements, such as changing its position; it is drowsy, drops objects placed in its hands, does not smile and does not even recognize its mother's voice. Voluntary or involuntary movements, produced, for instance, by painful irritations, are weak, the child being unable to sit upright or hold its head straight. The muscles of the limbs appear flaccid or spastic, and the child ultimately becomes paralyzed.

Simultaneously with changes in the general condition, visual disorders appear. The child seemingly does not notice objects placed before its eyes; it does not follow the light, does not notice its mother, and finally, at the end of a year, becomes totally blind. In contrast to the visual apathy, the acoustic and tactile impressions appear abnormally acute, the slightest noise startling the patient. This may be the cause of the convulsions frequently observed in such patients (Apert,[2] Higier,[3] Epstein[4]). The convulsions may be tonic and clonic, lasting one or two minutes, and may be provoked by strong painful irritations, whereas milder stimuli are usually without result.

In the terminal stage, the child becomes greatly emaciated, the head appearing too large in comparison with the rest of the body; the musculature is wasted and paralyzed; swallowing and sucking become impossible, and finally the child dies from inanition, after the disease has lasted from 15 to 18 months. A peculiar feature occurring in the latest stages is spasmodic laughter, such as is observed in pseudobulbar paralysis.

Objective Signs.—During the first stage of the disease, the general condition is practically normal, without visible changes in the size and configuration of the head or in the condition of the limbs. The child is motionless, the muscle power being retained and there are electrical degenerative changes. The tendon reflexes are usually exaggerated, with a positive Babinski sign, but they may be diminished or absent. The pupillary reflex can be elicited; but, as a rule it is sluggish. The cranial nerves show no change, except the optic atrophy combined with the typical changes first described in 1881 by an English ophthalmologist, Warren Tay:[5] "The optic disc, namely, appears quite healthy, but in the region of the yellow spot in each eye there is a conspicuous tolerably defined large white patch, more or less circular in outline, and showing at its center a brownish-red, fairly circular spot, contrasting strongly with the white patch surrounding it" (Fig. 41).

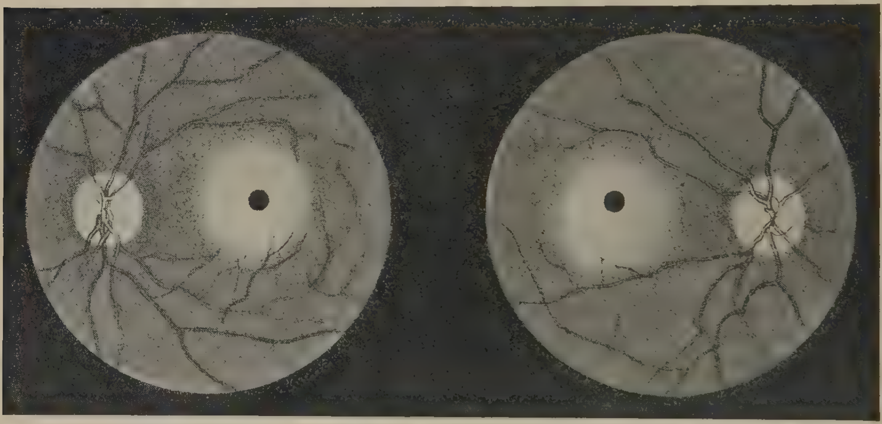

Fig. 41.—Disease in the region of the yellow spot in each eye, in an infant. (Warren Tay, in Trans. Ophth. Soc. of the United Kingdom, Vol. 1, 1881.)

The foregoing fundi changes—the red cherry spot surrounded by a white patch—are so characteristic that their absence prohibits the diagnosis of Tay-Sachs' disease being made, even when the clinical picture otherwise is typical (for instance, in the cases of Koller,[6] Patrick,[7] Kuh,[8] Mulberger[9]).

The clinical features outlined—gradually increasing apathy culminating in idiocy, inanition, progressive increase in visual disorders terminating in optic atrophy and the classical cherry spot—are supplemented by another typical feature, namely, the etiologic factor: it occurs exclusively in Jewish families coming from the former Russian Poland. Higier[10] of Warsaw pointed out the remarkable fact that in Poland, where one-sixth of the population is Jewish, amaurotic family idiocy (Tay-Sachs) is rather rare, the majority of cases having been observed in emigrants settled in other countries (America, Africa, and Australia). This racial proclivity to amaurotic family idiocy, though unexplained, remains the sole indisputable etiologic factor of this malady, as other causes, such as syphilis, alcoholism, infection, or tuberculosis, play no rôle whatever in its etiology. It is also a family disease, affecting children of the same family, though some may escape.

The **course** is always progressive, invariably ending in death, during the second year of the child's life.

Pathology.—The typical clinical features are combined with equally typical pathologic changes. These are found in the ganglion cells of the gray matter which are universally changed, in the brain, large basal ganglia, cerebellum, pons, medulla, spinal cord, spinal ganglia, retina, and even the sympathetic nervous system. The changes in the cerebral cortex are especially marked. The body of the ganglion cells is shaped like a balloon, bottle, or pear. The processes are swollen and well developed (Fig. 42). They show

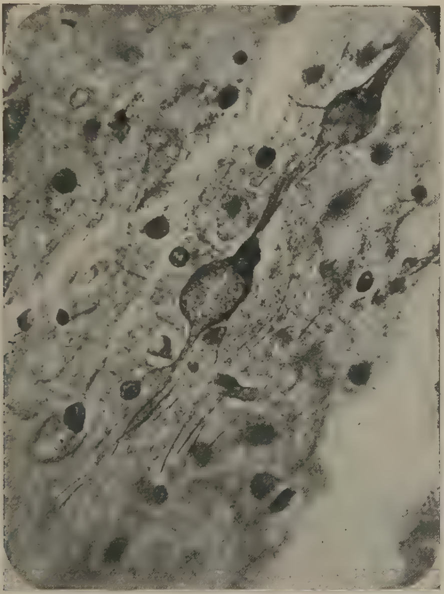

FIG. 42.—Photomicrograph of cortex of the frontal lobe in a case of amaurotic family idiocy. The pearlike shape of the ganglion cells, the swollen dendrons, the misplaced nuclei pushed into the apical dendrons, the fibrils surrounding the swollen cell bodies are well marked. The numerous dark round bodies scattered over the field are nuclei of glia cells and their modifications (gitter cells). Use a hand lens. (Bielschowsky's silver stain of a frozen section. × 450.)

excellently developed fibrils, which are also marked on the surface of the cell body (Fig. 43) in which the cytoplasm appears distinctly reticular or as a fine granular mass (Fig. 44) devoid of so-called Nissl bodies. The latter are gathered around the nucleus, which (Fig. 45) is always misplaced, appearing as if pushed into the apical dendron and surrounded by a well-developed membrane.

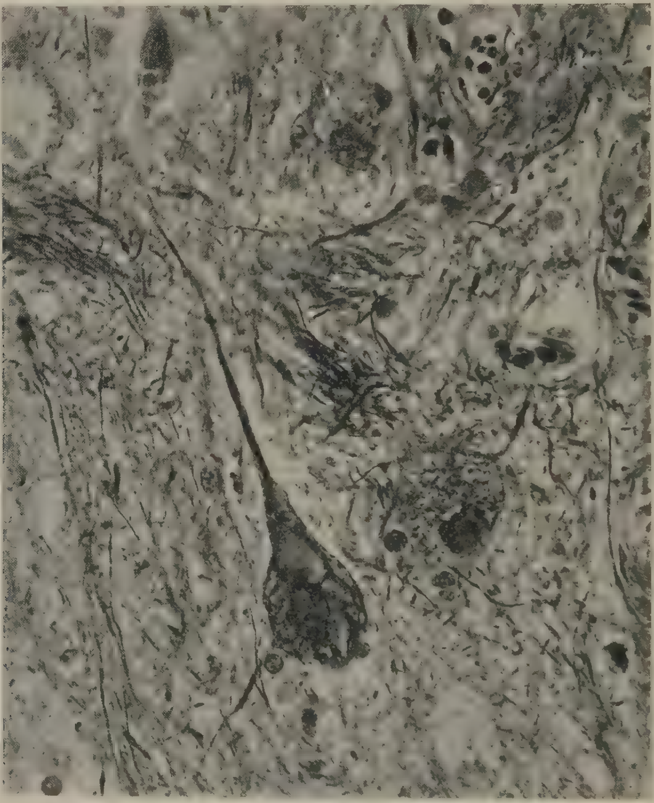

Fig. 43.—Ganglion cells of the ventral horns of the spinal cord (amaurotic family idiocy). The expanded cell bodies are distinctly reticulated. The reticulum in the cell to the left consists of long nerve fibrils covering the surface of the cell body and overlapping the so-called cytoplasmic reticulum better shown in the cell to the right. The rest of the picture shows numerous nerve fibers, glia cells, and blood vessels (in the upper right corner). (Bielschowsky's silver stain. × 560.)

It is noteworthy that, in spite of profound ganglion cell changes, there are none in the central nerve fibers, which sometimes show lack of myelinization. For instance, the pyramidal tract, the radiation fibers of Gratiolet, the occipital and temporal lobes, and other parts of the central nervous system, appeared in some cases poorly developed, their growth inhibited (Savini,[11] Naville[12]), but evidences of secondary degeneration were totally lacking. Many nerve fibers were represented merely by axons which did not have time to become medullated. In other words, many systems of fibers are in a state of aplasia, stop growing with the onset of the disease, which, as pointed out, sets in as early as the age of three months, and probably earlier. At this time, the eye changes are already present (Tay, Kingdon[13]). Another peculiarity is the absence of reactive changes in the glia, such as satellitosis and neuronaphagia. The glia cells in some places appear unusually large and rich in protoplasm, with an excellently developed chromatin-rich nucleus. In other places, such as the cortex of the brain, there is an apparent scarcity of glia cells, the visual field being represented by an enormous amount of densely crowded ganglia. Nor are there inflammatory changes in the vessels and the meninges. The former show vast accumulation of gitter cells in the adventitial spaces, the latter show, in addition, lymphocytes macrophages packed with pigment, etc.

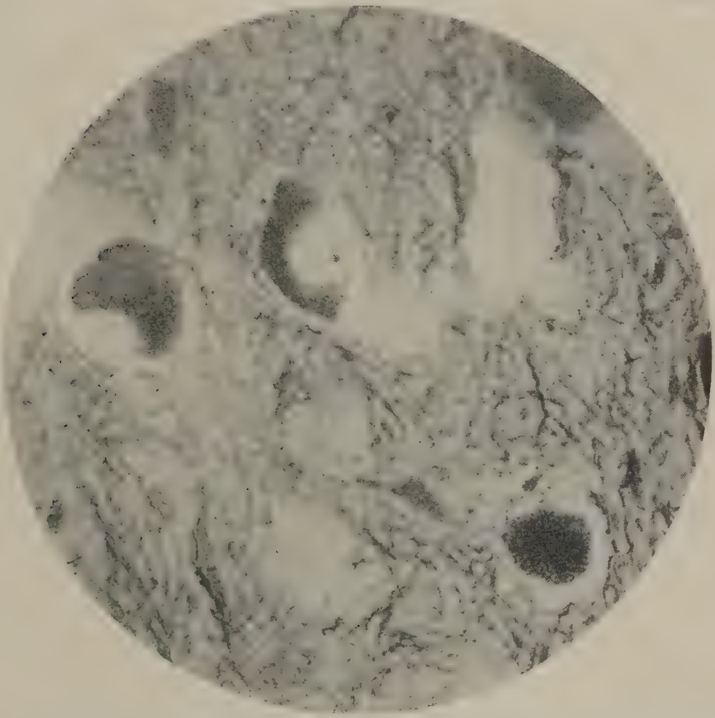

Fig. 44.—Ganglion cells of the ventral horns of the spinal cord. The larger portion of the cytoplasm is in the form of a granular mass. The smaller portion, around the nucleus, is pale and, when stained with toluidin blue, it shows a number of well-preserved Nissl bodies, as seen in Fig. 43. (Schaffer's hematoxylin staining method. × 200.)

The histopathologic studies, with the newest methods of staining, which cannot properly be considered here, leave no doubt that amaurotic idiocy, type Tay-Sachs, is both a developmental anomaly and a diffuse degeneration. The former shows as maldeveloped ganglion cells, capable of functioning, the latter follows later and manifests itself in destruction of the ganglion cells, especially of those of the optic thalamus.[14] The cause of both these conditions is to be looked for in some chemical, catabolic changes, or some embryonic defects, probably as Schaffer[15] suggested, of the ectodermic layer from which the ganglion and glia cells arise. Talbot,[16] studying the metabolism in his case, found it to be very low, approximately 30 per cent. lower than in three normal children of the same weight and age. These findings are of great importance. Further studies along biochemic and histopathologic lines will undoubtedly bring out new facts which will enable us to understand the nature of this remarkable disease.

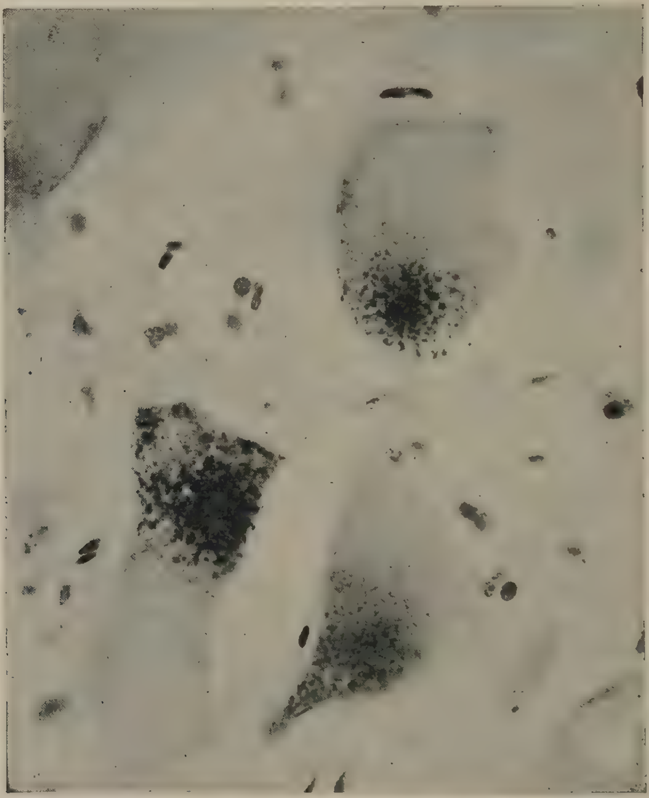

FIG. 45.—Spinal cord cells. The mass of dark granules are Nissl bodies in an excellent state of preservation. (Toluidin blue. × 500.)

Treatment can, of course, be only symptomatic.

Type Vogt.[17]—This is the juvenile type of amaurotic family idiocy. The cases belonging to this type also come, though not exclusively, from Jewish families. Children of the same family born healthy may show no abnormalities up to the age of six or later (even up to 14 years). At the latter age, they begin to show visual disorders, which in a few months end in complete blindness and optic atrophy. Further mental development ceases and gradually declines; the patient loses his knowledge of reading and writing, and even the faculty of speech; he becomes unsocial, untidy, apathetic to his surroundings, relatives and even to his mother. The intellectual decay proceeds simultaneously with the physical decline; muscular weakness, which first shows in the back and the limbs, gradually develops into paralysis (flaccid or spastic), with lost or exaggerated reflexes. The growth of the body stops; marasmus sets in gradually, and, after a protracted course, leads to death. As to etiology, nothing is known, except that some families show a predisposition to this disease. As in the previous group, syphilis, tuberculosis, and alcoholism play no rôle.

Pathologically, the cases of Vogt are almost exactly like those of Tay-Sachs, and Vogt comes to the conclusion that his type of amaurotic family idiocy represents but a later stage of the classical form of Tay-Sachs, which is termed infantile. Ichikawa,[18] Batten,[19] and others arrived at the same conclusion. Somewhat similar cases have been reported in the older literature by Pelizaeus,[20] who could trace the outlined clinical picture, except the presence of mental symptoms, through three generations. Higier[21] reported a similar syndrome in four sisters (12 years, 10 years, 9 years and 7 years) in whom the mental symptoms and optic atrophy were quite pronounced. However, neither Pelizaeus, nor Higier's cases belong to this disease as justly pointed out by Merzlacker.[22]

Spielmeyer's Type.[23]—Spielmeyer records a case of amaurotic idiocy in a family of five children. The oldest child was healthy. After its birth the father contracted syphilis. The following four children each became ill at the age of six years, with epileptic seizures, rapidly developing dementia and blindness. Death followed after a course of from 6 to 10 years. These differ from the Sachs type in the onset at the age of six, the protracted course, absence of macular changes and a few other minor differences. Similar features consisted in the familial occurrence, early onset of dementia and blindness. Differences are shown also in the pathologic changes. In Spielmeyer's cases, the ganglion cells preserved their configuration, though they were slightly swollen, the dendrons being normal. The cell bodies contained large amounts of greenish-yellow pigment, but not the fat-life (lipoid) substances which are sometimes present in amaurotic family idiocy (Sachs and Strauss,[21] personal observation). Spielmeyer's cases are generally referred to as belonging to the juvenile type of Vogt, but, in view of the specific etiology and the peculiar pathologic findings, Spielmeyer's cases should, at least at present, be placed in a group by themselves.

Late-infantile type—especially well studied by Bielschowsky.[25] In this type the disease begins at the age of four with practically the same symptoms as in the infantile form, but without the characteristic macular changes. Optic atrophy and insular atrophy of the retina are present; this type is not confined to the Jewish race and lasts much longer (three to four years).

Pathologically it is similar to the classical type of Sachs, but here marked atrophy of the cerebellum, with absense of its granular layer, mossy, tangential and basket fibers, is present. The dendrons are less involved, and many ganglion cells in the subcortical areas are quite normal. Many other cases have been described in which the fundamental features—familial occurrence, blindness, and idiocy—were quite pronounced, while other equally important signs, such as macular changes and a specific etiology, were lacking. In some cases, for instance in the one recorded by Mayou, both infantile and juvenile types were combined. In this case, three healthy children from a consanguineous marriage began to show, at the age of seven, a decline of intelligence and vision. The retina was irregularly covered with granular pigment and showed the typical red cherry spot.

On the other hand, Winkler and Wan West's[27] case demonstrates that some forms of amaurotic idiocy may be purely congenital. In their case, two of six children (not Jewish) exhibited identical symptoms. The history of one (five years old) is given in detail. It was born with a small head and blind. From the age of two months, it suffered from repeated convulsions (about 25 attacks a day); at the age of eight months, contractures of the extremities set in. During the last year of life, bulbar symptoms developed, followed by marasmus and death at the age of 5½ years. The child was blind, deaf, and spastic; the optic discs were white, without macular changes.

The post-mortem examination revealed a defective and asymmetric brain (deficient occipital lobes, absence of corpus callosum and of geniculate bodies), the hemispheres resembled those of a seven or eight months' old fetus. The histologic examination showed ganglion cells increased in size, but their number was diminished, and there were some vascular changes. Another child of the same family exhibited similar symptoms: Microcephaly, blindness, idiocy, and atrophy, with contractures of the extremities.

It is needless to insist that Winkler's case has nothing to do with the classical type of amaurotic family idiocy. It merely shows that many types of idiots become blind, but the combination of idiocy, blindness, and familial features does not justify diagnosing them as amaurotic family idiocy in the sense in which Sachs described this morbid condition. An exception should be made for the so-called juvenile type of Vogt and the late infantile types, yet even these forms are by far too rare to be considered as specific clinical entities. On the contrary as Globus[28] has shown the three types of amaurotic family idiocy—infantile, juvenile and late infantile—clinically and pathologically have so much in common that a sharp line between them cannot be drawn.

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PALSIES OF PERIPHERAL NERVES AND PLEXUSES

(A) TRAUMATIC

A peripheral nerve trunk is composed of a number of bundles (fasciculi) surrounded by a powerful connective tissue membrane—epineurium, which not only envelops but also penetrates the nerve trunk, dividing and separating its bundles from one another (Fig. 46). The epineural membrane contains an abundance of fat, elastic tissue, and vessels. Each of the bundles making up the nerve is enveloped by a distinct connective tissue membrane—perineurium, which gives off prolongations dividing the bundles into smaller ones (funiculi). These intrafunicular prolongations are called endoneurium by Key and Retzius.[1] The three coverings of the nerve fibers—epineurium, perineurium and endoneurium—are derived from the meninges of the brain and spinal cord. Key and Retzius[1] definitely assert the analogy between the perineurium and the arachnoid membrane, of which the former is the direct prolongation. They also definitely established the prolongations of the subdural and subarachnoid spaces of the brain

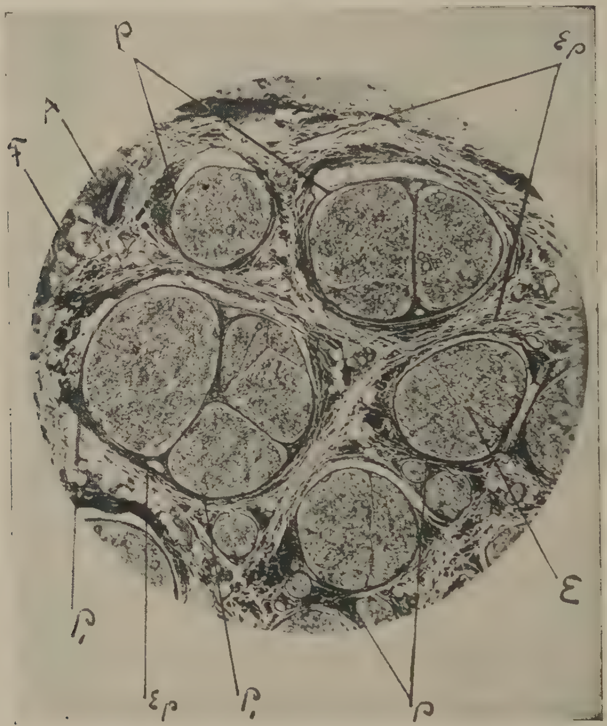

FIG. 46.—Photomicrograph of a portion of a transversely cut peripheral nerve: *Ep*, epineurium containing fat (*F*) and blood vessels; *A*, artery; *P*, perineurium separated from the fasciculus by a white zone (*P*) which in all probability is a distinct membrane; *E*, endoneurium dividing the fasciculus into funiculi. (Alzheimer-Mann stain of a celloidin section 15 microns thick. × 45.)

and spinal cord into the peripheral nerves; consequently they possess the same spaces as the brain and spinal cord. The perineurium, like the epineurium, carries blood-vessels; the endoneurium, lymphatics. The nerve fiber itself consists of a Schwann's membrane made up of cells containing an oblong nucleus, rather poor in chromatin, surrounded, at its poles, by a granular cytoplasm. The nucleus is situated inside the Schwann's membrane, between the latter and the myelin sheath.

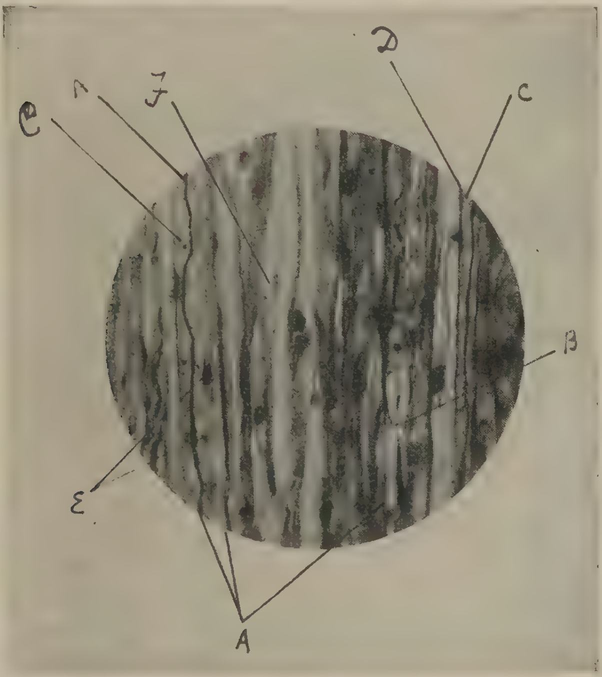

Fig. 47.—Photomicrograph of longitudinal section of a normal peripheral nerve: *A*, axons; *B*, Ranvier crossing; *C*, myelin sheath; *D*, Schwann's membrane; *E*, Schwann nuclei; *F*, nucleus of connective tissue (endoneurium). Use a hand lens. (Bielschowsky-Alzheimer-Mann stain of a celloidin section, 15 microns thick. × 350.)

Morphologically and physiologically, the Schwann cells are identical with the glia cells, being looked upon as peripheral glia. At certain intervals, the Schwann membrane forms constrictions, known as Ranvier crossings (Fig. 47), where it comes in contact with the central portion of the nerve—the axon. The interval between two crossings contains a nucleus, and the resulting segment may be looked upon as a cell. In other words, the Schwann membrane is distinctly segmentary in its structure. In the central nerve fibers (brain, medulla, spinal cord) the Schwann's membrane is lacking. Close to the Schwann's membrane, and separated from it by the nuclei, is the myelin sheath (Fig. 47). This is homogeneous and divided by so-called Lantermann's incisions into segments. The Lantermann's incisions are considered by Key and Retzius to be artefacts and, by the latest authors, to be original formations. (In specimens stained by the Alzheimer-Mann or by a combined Bielschowsky-Alzheimer-Mann method, I could not see these incisions.)

The center of the nerve fiber is occupied by its most important constituent—the axis cylinder or axon (Fig. 47). The latter, fibrillary in structure, consists of delicate, so-called primitive fibrils, separated from one another by a homogeneous plasmatic substance—the axoplasm. Reaching the muscle fiber, the axon splits up in non-medullated fibrils, terminating as a fibrillary network. The myelin sheath is present in every central nerve fiber, but it is absent in the majority of the sympathetic nerves and in some peripheral nerves. Ranson[2] asserts that the latter are preferably made up of non-medullated fibers.

In newborn and very young infants, under five weeks of age, the peripheral nerves show some structural peculiarities. Thus C. Westphal[3] and A. Westphal[4] found the myelin poorly developed, very thin, or broken up in fragments, stainable with osmic acid, not in a dark, but in a greenish or yellowish color. Many axons are unusually large, often totally deprived of myelin and without Ranvier's crossings and Lantermann's incisions. The Schwann cells, on the contrary, are very numerous and, like the endoperineurium, well developed. During the sixth week of life the myelin is quite fully developed. A. Westphal is of the opinion that the noted anatomic features are responsible for the electrical peculiarities to be considered elsewhere.

The *chief* physiologic quality of a nerve is the degeneration of the portion separated from the center, that is from the ganglion cell. The resulting degenerative changes are known as secondary or Wallerian, having first been described by Waller. They are sometimes in evidence within 24 hours after the nerve has been divided. The myelin and axon break up into fragments, forming myelin globules, drops, and droplets. The broken-up fragments are picked up by the glia (Schwann) cells, transformed into lipoid, fat-like substances, and removed to the blood-vessels for final elimination. Simultaneously, there is a great proliferation of the Schwann cells, which form long rows or bands of fibers, along the course of which the new nerve fibers grow, leading to the regeneration of the injured nerve, the regenerative process always arising from the central stump. Under unfavorable conditions, the glia is replaced by connective tissue. Then the nerve perishes, becoming transformed into, or replaced by, a connective tissue strand or scar. Regeneration of the nerve is, in such a case, possible only when the scar is removed.

The degeneration of a nerve is followed by that of the muscle, which becomes atrophied and finally replaced by connective tissue.

The changes, as outlined, occur in a nerve that is totally severed (cut, torn, lacerated). In cases of incomplete injury, caused, for instance, by pressure (from tumors, hemorrhages, bone fragments, foreign bodies, etc.), some of the nerve fibers may escape injury, and the clinical picture will be somewhat different from that in a completely cut nerve.

Symptoms.—As a peripheral nerve contains both motor and sensory fibers, the symptoms of nerve injury are motor (paralysis, paresis) and sensory, combined with electrical, vasomotor, and trophic disturbances.

Motor symptoms constitute a complete (paralysis) or incomplete (paresis) loss of voluntary movements. The paralysis is associated with loss of muscle tone and reflexes, atrophy of the corresponding muscles, and electrical changes. The affected muscles are flabby and soft, and exhibit no resistance to passive movements, while the active or voluntary movements are absent. In short, the paralysis is flaccid, degenerative, and atrophic. The degenerative features can be determined from the electrical changes in the nerves and muscles. In severe cases, within from 12 to 14 days after the nerve injury, the faradic current fails to elicit any response from either the nerve or the muscles, while the galvanic current will cause a muscular contraction, provided it is applied to the muscle direct, but not when it is applied to the nerve. In other words, the direct galvanic response is retained or even increased, while the indirect (applied to the nerve) is absent. The galvanic direct muscular contractions, however, though readily obtainable, are by no means normal, being flabby and worm-like, with the anodal closure contraction (A.C.C.) stronger than the cathodal closure contraction (C.C.C.). This is a reversal of the normal formula, and is called the reaction of degeneration (R.D.). In less severe injuries, the reaction of degeneration is but partial, occurring in various forms. For instance, both the muscle and the nerve may respond to the faradic current, while the galvanic may give an inverted polar formula, or a worm-like muscular contraction; or there may be slight or no response to the faradic current, the galvanic eliciting practically normal electrical reaction. In other cases the electrical changes are quantitative, a much stronger current being required to elicit a muscular contraction, the polar formula remaining unchanged.

While the foregoing considerations are true for a grown person, in newborn and very young infants the electrical conditions are somewhat different.

As shown by the Westphals, in newborn children and young infants up to five weeks of age, the faradic as well as the galvanic excitability of the nerves and muscles is diminished *quantitatively*, much stronger currents being required to evoke a contraction than in adults. This is true for both cranial and spinal nerves. Currents producing in adults a marked muscular contraction are without effect when applied to the same points in a newborn infant, and very strong currents produce only weak contractions. The latter, slow in coming and slow in disappearing, are not accompanied with any signs of discomfort or pain. In other words, young infants easily stand strong currents, 10 milliamperes, for instance, even when they are applied to the face. A. Westphal, however, adds that *direct* galvanic excitability, even in the youngest infants, is practically normal. He explains the foregoing electrical peculiarities by the deficient myelin development in the nerve fibers of the newborn.

Sensory disturbances are comparatively less in evidence, or they may be temporarily present, or entirely absent. When present, as a rule, they involve a territory much smaller than that indicated anatomically or physiologically. Generally speaking, the loss or decrease in sensation (anesthesia, hyperesthesia) pertains to a portion of an area innervated by the injured nerve. It is of the so-called *peripheral* type, and involves usually all sensations alike—pain, temperature, touch, pressure. The fact that in some cases of peripheral lesions sensory disturbances may be lacking, may rapidly disappear, or may occupy but a very small area is explained by (*a*) the presence of anastomoses between the peripheral nerves, and (*b*) the *collateral* innervation through neighboring or adjacent nerves, (*c*) the overlapping of sensory areas, (*d*) so-called "protopathic" and "epicritic" sensibilities, as described by Head, and some other factors. According to Head and his school, sensory peripheral impulses are conveyed by three separate systems of fibers: Deep, protopathic, and epicritic. To the *deep* he refers pressure, sense of motion, position, and vibration; to the *protopathic*, pain, temperature below 20°C. and above 40°C. (extreme cold and heat); to the *epicritic*, moderate temperature (between 20 and 40°C.), fine localization sense, and touch (with cotton wool). The deep sensibility is, according to this teaching, conveyed by subcutaneous nerve fibers terminating in the tendons, fascia and joints, while the epicritic and protopathic are conducted by nerve fibers situated in the skin proper. When a nerve is in process of regeneration, the epicritic sensibility occupies a larger area and returns much later than protopathic sensibility. However, the early return of the protopathic sensation (pin-prick), according to L. J. Pollock,[5] is not caused by regeneration of problematic specific nerve fibers, but is due to the fact that there is overlapping of innervation in the affected region. For instance, the musculospiral nerve supplies a region on the skin which is also innervated by the median and musculocutaneous nerves. When the musculospiral nerve is cut, only the area supplied by it will be involved in the anesthesia, while the portions supplied by the other two nerves will not be so involved. This is the reason that the anesthesia usually occupies a smaller area (the area of exclusive nerve supply) than is indicated by the anatomic distribution of the nerve. The remainder of the area is supplied by the other two nerves, which are not damaged. When in the cut musculospiral nerve, for instance, the sensibility to prick returns before the sense of touch, it is not because of the dissociation of epicritic and protopathic sensibilities, but because of the assumption of function by the neighboring, overlapping nerves which were not damaged. When the overlapping nerves are injured or cut, the anesthesia occupies a much larger area. Pollock,[5] Trotter, Davis, Stookey, Boring and Cobb[6] are of the opinion that the areas described by Head merely represent varying grades of hyperesthesia, and that the use of cotton wool for determining the sense of touch, and of pin-pricks for determining the sense of pain are sufficient clinically, as the areas of the former usually coincide with the latter. In pediatric practice Head's ideas and classification are of no value, and are not used by the majority of clinicians.

Vasomotor disturbances (redness and cyanosis of skin, burning, and edema) and *trophic* changes (herpes, glossy skin, pemphigus, bone atrophy, lack of nail and hair growth, etc.) are usually absent when a nerve is completely severed. They occur mostly in *incomplete* nerve lesions. In the latter, abnormal sensations are frequently present—*paresthesias*, such as pin, needle, heat and cold sensations, and burning.

The **prognosis** and **course** of traumatic nerve injuries depend upon the severity of the anatomic lesion, its duration, etiologic factors, electrical changes, and sometimes on the treatment. As a rule, a damaged peripheral nerve will recover completely without any active treatment, the recovery being accomplished through the growth of the central stump connected with the nerve cell. This is at present almost generally accepted as a fact. According to Elsberg,[7] the average time before the return of the first voluntary movements is six months for the median nerve; eight months for the musculospiral; ten for the external popliteal; eleven for the internal popliteal and ulnar nerves, and sixteen for the sciatic nerve. A compressed nerve usually recovers its function in about thirty days. Absence of electrical changes within from twelve to fourteen days after injury makes the prognosis favorable, and recovery may be expected within a week. Partial reaction of degeneration indicates a delayed recovery, from six to eight weeks, while in the presence of the complete reaction of degeneration recovery cannot be expected before six months. If neither the nerves nor the muscles respond to electrical stimulation, the prognosis is absolutely unfavorable; recovery cannot be expected without proper treatment.

Treatment.—In mild cases of nerve injuries, caused, for instance, by pressure, active interference is unnecessary. Rest and symptomatic treatment, such as heat applications for relief of pain, are sufficient. With the passing of the acute stage, massage, exercises, or the faradic or galvanic current may be used. If nerves are cut, surgical treatment is necessary. End-to-end suture has proved to be the best procedure. Neurolysis, that is, freeing the nerve from a body to which it adheres (bones, scars, tumors), or in which it is embedded, is often beneficial. Large gaps must be refilled or bridged by transplants, the so-called autotransplants or autogenous transplants being the best. A cutaneous branch, such as one of the musculocutaneous or musculospiral nerves, is excised, and sutured to the freshened ends of the peripheral and central stumps. Anastomoses and implantations are useless and have been abandoned. In milder cases no surgical treatment is necessary, the results being almost as good as with surgical interference.

(B) Non-traumatic Isolated Nerve and Plexus Paralyses

An isolated paralysis of a nerve or plexus may result from other than traumatic causes. Thus, intoxications (alcohol, arsenic, lead), infections (diphtheria, typhoid fever, tuberculosis, erysipelas), or chemical irritations (hypodermic injections of ether, alcohol) may produce changes in one or more peripheral nerves, changes commonly classified as neuritis. In this classification also belong nerve lesions caused by changes in a neighboring organ, such as pressure by a bone (exostoses, cervical rib, tumor), infection (caries of spine, mastoid, appendicitis), and joint lesion (articular neuritis). The joint lesion may have sprung from a contusion, slight, but sufficient to produce a neuritis, without apparent cause unless the history of injury can be elicited. Finally, there is so-called professional neuritis, rather unusual in children, and ascending neuritis. The latter originates in a nerve wound which, though healed, may be followed by a neuritis which spreads upward. According to some authors, this is due to an infection which passes along the lymphatics of the nerve, being frequently associated with hysteria (Moty[8]).

It is customary to classify the inflammatory nerve changes as parenchymatous and interstitial neuritis. In the former, the parenchyma of the nerve (axon and myelin), in the latter, the connective tissue membranes (endoneurium, perineurium, and epineurium) are principally involved. As a rule, however, both the parenchyma and the interstitial tissue are more or less involved, and pathologically the differences are only quantitative. The parenchymatous changes show early in the myelin sheath, which breaks up into fragments, globules, drops, and droplets. This is followed by proliferation of the Schwann cells, many of which contain myelin fragments. The destruction of the myelin is not confined to the entire length of the nerve fibers, but usually proceeds in segments, some of which show an apparently healthy myelin, some appear naked, that is, deprived of the myelin, while in other segments the myelin is broken up. This is the segmentary type of neuritis (neuritis peri-axialis of Gombault). The axon may appear slightly granular or may long remain unchanged. In the latest stages, the axon becomes irregular in its course, shows thickened and thinned portions, and finally dies, showing the Wallerian degeneration which is the last stage. The nerve fiber shows, in this condition, only the Schwann membrane, and consists of the oval nuclei and long protoplasmatic processes forming bands of fibers. Still, when refined staining methods are used, such as Bielschowsky's silver stain, even in this apparently empty membrane, a large number of thin nerve fibers, axons, may be seen, occasionally invested with a delicate myelin sheath. These young thin fibrils are regenerated fibers. In neuritis these can always be found by the side of degenerated fibers. The simultaneous presence of normal degenerated and regenerated fibers is a characteristic feature of a neuritic process.

The interstitial phenomena are confined to vascular and proliferative changes. The former are hyperemia and infiltration of the smaller vessels with lymphocytes, polyblasts, and granular or fat granule cells. In some cases, hemorrhages occur between the nerve fibers. In chronic cases, the connective tissue cells may proliferate and lead to thickenings in the nerve.

Symptoms.—As in the traumatic nerve lesions, the symptoms are motor, sensory, trophic and vasomotor, usually resembling those noted in conditions in which the nerve is incompletely destroyed or cut. The most frequent symptom in this condition, as well as in neuritis, is pain, especially marked in interstitial neuritis. The pain is spontaneous, constant, confined to, and radiating along, the area supplied by the nerve. It increases on active or passive movement, and especially when pressure is applied. Touch may be painful; it is then called hyperesthesia. In milder cases, there may be merely paresthesias, that is, abnormal painful sensations, such as burning pin-and-needle sensations and feeling of numbness. In long-standing or advanced cases, the involved area may show diminished sensation (hypesthesia), or may be anesthetic, insensible to pin-pricks.

The same motor disturbances (paresis, paralysis), loss of reflexes, trophic lesions, electrical changes occur as in traumatic nerve lesions.

The **prognosis** and **course** depend essentially on the etiology. They are good if a nerve can be released from pressure, or an infection cleared up, for, as the histopathologic studies show, even a badly damaged nerve contains a great number of young, regenerating fibers.

Treatment.—In the acute stage, rest with the affected limb in proper position, and hot applications, such as poultices or heated air, may be all that are necessary. In some cases salicylates and especially pyramidon afford relief. Morphine, codein, and other opiates may be used in case the suffering is very severe, in the initial period. When the acute stage has passed, that is, when the pain subsides, massage or electrical treatment may be used. The use of the faradic or galvanic current depends on the condition of the nerve or muscle. If it does not respond to the faradic current, the galvanic, of course, should be employed.

The etiology, symptomatology, course, and treatment of traumatic and non-traumatic nerve lesions will be considered in somewhat greater detail in the next chapters, in which the diseases of single nerves will be discussed. The cranial nerves will be considered first, except the first, second, and eighth (olfactory, optic, and acoustic), as strictly speaking, they are not peripheral nerves.

Third Nerve, Nervus Oculomotorius.—This nerve arises in several bundles (fasciculi) from cell bodies forming seven distinct nuclei. The latter are situated beneath the aqueduct of Sylvius, in the region of the anterior corpora quadrigemina, dorsad to the posterior longitudinal bundle. The fasciculi appear in the base of the brain in the form of a single nerve trunk, at the inner side of the cerebral crura, in the so-called trigonum interpenduculare, situated in the posterior skull cavity. From the latter it enters the middle skull cavity and pierces the dura at the level of the anterior and posterior clinoid processes of the sphenoid bone. Then it enters the outer wall of the cavernous sinus, running toward the sphenoidal fissure, where it divides into superior and inferior branches, the former of which supplies the superior rectus and levator palpebræ superioris; the latter, the rectus internus, rectus inferior, oblique inferior, the ciliary (accommodation) and sphincter iridis (pupillary) muscles.

The most important structures, therefore, that the oculomotor nerve touches, on its journey from the brain to the muscles, are: The cerebral crura, dura mater, sphenoid bone, cavernous sinus, sphenoid fissure, as well as the posterior and middle skull cavities. It follows that abnormalities in these regions, such as tumors, hemorrhages, fractures of basilar skull bones, dura lesions (tumors, syphilis), and sinus troubles, may indirectly involve the third nerve. In fact, diseases of its neighboring structures most commonly cause third nerve paralysis. Syphilitic involvement of the meninges is most frequent (such as syphilitic leptomeningitis or pachymeningitis, syphilitic periostitis and gumma). Other affections, as sinus troubles or skull lesions, are less frequent.

Various *infections* follow syphilis, in the order of frequency, as causes of oculomotor paralysis. Of these, diphtheria stands first. The other infectious diseases (scarlet fever, measles, grip, rheumatism), intoxications (alcohol, ptomain poisoning), and constitutional diseases (diabetes, for instance) are causative in a much smaller percentage of cases.

Symptoms.—As the third nerve is a purely motor nerve, the symptoms will be motor, exhibited in paralysis of the muscles supplied by this nerve. Thus, paralysis of the levator palpebræ superioris will cause drooping of the upper lid (ptosis) and inability to open the affected eye voluntarily. Paralysis of the superior rectus muscle will cause inability to move the eye-ball upward; in paralysis of the internal rectus muscle, the eye will be turned outward, resulting in an external or divergent strabismus. Oblique inferior paralysis will cause inability to turn the eye upward and somewhat outward. Paralysis of the pupillary sphincter will lead to the dilatation of the pupil and its inability to react to light, while paralysis of the ciliary muscles will result in loss of accommodation. On account of the weakness of the ocular muscles, the eye-ball will protrude, resulting in slight exophthalmos. The results of oculomotor paralysis, therefore, are: internal ophthalmoplegia (loss of pupillary reaction to light and accommodation), partial external ophthalmoplegia, strabismus, ptosis, and exophthalmos. Subjective complaints may be absent, except possibly double vision (diplopia) if the patient raises the drooping upper lid and tries to look with both eyes.

A *peripheral* third nerve paralysis is shown by total paralysis of all the muscles it supplies (extrinsic and intrinsic). If the lesion is intracerebral, the paralysis is *partial* and indicates a central or nuclear involvement. For instance, an isolated paralysis of the pupil, or of some extrinsic muscle, as the levator palpebræ superioris, usually indicates brain lesion proper (central or nuclear lesion).

The **prognosis, course and treatment** depend upon the original cause. If it is a tumor pressing on the nerve or a fractured bone, the prognosis is bad. As the cause is usually syphilitic, the prognosis is comparatively good, especially if an early diagnosis is made, followed by the appropriate treatment. The paralysis sometimes recedes within a few days, and remarkable improvement is shown in the general condition of the patient.

Periodic Oculomotor Paralysis.—This condition has been noted occasionally in children. Of 74 cases collected by Flatau[9] 11 occurred in children between 11 and 15 years of age, two at from 6 to 10 years, and one before five. As the name indicates, the oculomotor paralysis sets in at more or less regular intervals, preceded by an ipsilateral headache and vomiting. Both usually disappear with the onset of the paralysis. The latter may last for days, weeks, or months, may disappear totally or partially, may recur with the same symptoms and run an identical course. The headache and vomiting in their periodicity closely resemble migraine. This led Charcot, in 1890, to call these migrainous symptoms, accompanied or followed by oculomotor paralysis, ophthalmoplegic migraine, a view sustained by subsequent observations. The paralysis, ordinarily total, is confined to the side on which the headache occurs and affects both extrinsic and intrinsic eye muscles supplied by the third nerve. The fourth and sixth nerves may also be affected. In some cases, as shown by Ballet and others, fever is present. In a few cases the paralysis is partial, or the total paralysis develops gradually. The paralysis disappears slowly, within days or weeks. A free interval follows in which some remnants of paralysis may be present. The intervals last for variable periods. The cause of periodic oculomotor paralysis is not known. Since paralysis of the typical peripheral type may be caused by infection or intoxication or, as pointed out in the previous chapter, by involvement of neighboring organs (dura, sphenoid bone and cavernous sinus lesions, orbital bone involvement, etc.), the cavernous sinus might hold the secret of a cause of the recurrent paralysis of the third nerve. That this is possible is evidenced by the severe chronic periodic headaches, the occasional elevation of temperature, occasional involvement of the fourth nerve, which passes this sinus, and other factors mentioned in the chapter on migraine. Möbius pointed out that frontal sinus trouble might be a possible cause of recurrent ophthalmoplegia. In his case the attack lasted four weeks, disappearing after a violent attack of sneezing.

The **fourth nerve,** nervus trochlearis, also originates beneath the aqueduct of Sylvius, but in the region of the posterior corpora quadrigemina. It appears on the base of the brain at the anterior margin of the pons and on the external surface of the crura around which it winds. It pierces the dura near the posterior clinoid processes of the sphenoid bone, runs forward in the outer wall of the cavernous

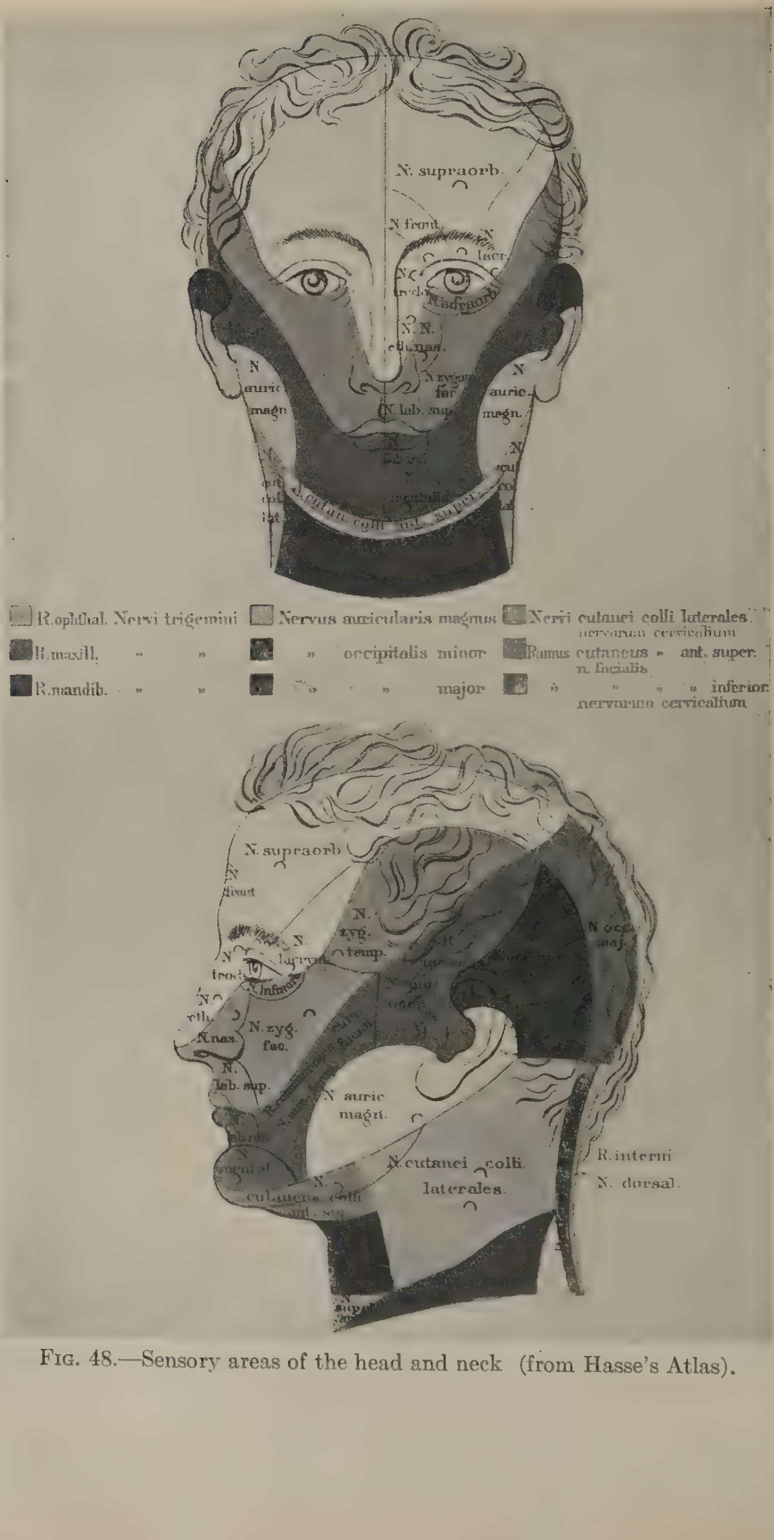

FIG. 48.—Sensory areas of the head and neck (from Hasse's Atlas).

sinus, and passes through the inner part of the sphenoidal fissure, ending in the superior oblique muscle.

The causes of paralysis of this nerve are similar to those of paralysis of the third nerve, the organs through which it passes being usually involved. The main action of the superior oblique nerve is to turn the eye down and inward. Paralysis of this nerve will cause the rolling of the eye upward and somewhat toward the healthy side. There will be difficulty in turning the eye downward, as in walking upstairs or downstairs. In the lower half of the visual field there will be double images. As paralysis of this nerve rarely occurs, it is of little clinical value.

The **fifth nerve (nervus trigeminus)** is a mixed nerve, containing motor and sensory fibers. The motor fibers originate in a nucleus situated in the so-called segmental portion of the pons, in its upper and lateral region. The larger portion of the fifth nerve is made up of the sensory fibers, originating in nerve cells of the gasserian ganglion situated on the pyramidal bone. Both motor and sensory fibers pierce the dura, leaving the skull through the foramen ovale (motor and sensory branches) and foramen rotundum (sensory branch). Another sensory branch (the ophthalmic), before leaving the skull cavity, enters the orbit through the sphenoidal fissure, running partly in the external wall of the cavernous sinus. Each of the three sensory divisions of the fifth nerve has definite cutaneous sensory areas. Thus the ophthalmic branch (first division) supplies the forehead, anterior portion of the scalp, the upper eyelid, the outer surface of the nose, the glabella, the septum and turbinated bones of the nose, its mucosa, the conjunctiva and cornea; the second division, the maxillary, enters the orbit through the sphenomaxillary fissure, supplying the upper lip, nose, lower eyelid, cheek, its mucous membrane, and the upper teeth. The sensory portion of the third branch (mandibular) supplies the dura, external auditory meatus, tragus of the ear, the chin, lower lip and its mucosa, lower teeth and gums. The sensory distributions are well shown in Fig. 48.

The motor fibers of the mandibular nerve innervate the muscles of mastication: The temporal, masseter, external pterygoid and myelohyoid, and the anterior branch of the digastric.

Etiology.—The etiologic factors (injuries, intoxications, etc.) in third and fourth nerve lesions will also cause paralysis of the fifth nerve. Fractures of the base of the skull, pons lesions (tumor), traumas of the orbital bones, of the bones of the face, may lead to fifth nerve paralysis.

Symptoms.—Though isolated lesions of this nerve are rare, in some cases its involvement, especially of the sensory portion, may be the early and sole sign of a cerebral lesion. For instance, a lost corneal or conjunctival reflex is sometimes characteristic of tumors of the cerebellopontine angle, a fact established by Oppenheim and confirmed by numerous observers. Other symptoms are mainly sensory, confined to the areas of distribution of the various branches.

Anesthesia of these areas as well as of the conjunctiva, or of the nasal mucosa, the loss of some mucous reflexes (nasal, conjunctival) indicate a fifth nerve lesion. The motor symptoms are evidenced by paralysis of muscles of mastication—opening or closing of the mouth. The lateral movements are deficient and performed without force. The muscles are soft to touch, flabby and weak; biting, for instance, is done with little or no force. In unilateral paralysis of this nerve, lateral movements are possible only toward the diseased side, the jaw being deviated toward it.

In addition to the motor and sensory disturbances, trophic lesions are often observed in the cornea, which becomes opaque and ulcerated, the bulbus becoming inflamed (keratitis).

Prognosis, course, and treatment depend on the etiology.

Sixth Nerve.—This nerve originates in a nucleus situated in the tegmental region of the pons, which it passes in emerging on the base, usually between the pons and the pyramids. Before it enters the orbit, this nerve pierces the dura, enters the floor of the cavernous sinus and the sphenoidal fissure. During its course, the sixth nerve comes in contact with the basilar artery, sphenoid bone, petrous portion of the temporal bone, and the carotid artery. It supplies the external rectus muscle.

Etiology.—A lesion of the pons, meninges, cavernous sinuses or other structures with which the nerve comes in contact may cause its paralysis. Tumors of the brain not pressing directly on the nerve, such as cerebellopontine tumors, may bring on a paralysis from strangulation by an underlying vessel (Cushing). Toxic and infectious causes play the same rôle here as in oculomotor nerve lesions.

Symptoms.—These are purely motor, confined to paralysis of the external muscle of the eye. The latter cannot move outward, but will be drawn inward, resulting in strabismus and diplopia (double vision).

The **course, prognosis,** and **treatment** are the same as for the nerves previously discussed.

The **seventh nerve (nervus facialis)** starts from a nucleus in the tegmental (dorsal) region of the pons where its fibers, before leaving the latter, wind around the sixth nerve nucleus and run upward and downward, forming ascending and descending branches. Leaving the pons and piercing the meninges, the nerve, in company with the acoustic nerve, enters the internal auditory meatus (porus acusticus), passes into the Fallopian canal, and emerges through the stylomastoid, foramen. It branches off on the face into numerous anastomosing twigs, which form a plexus known as pes anserinus.

While in the Fallopian canal, the facial nerve gives up six branches, of which the most important, clinically, are the chorda tympani, nervus stapedius, and Wrisberg's nerve. Other branches (nervus petrosus superficialis major, nervus petrosus superficialis minor and external petrosus) connect the facial nerve through the geniculate ganglion with the sympathetic ganglions (otic and Meckel's).

The chorda tympani starts in the geniculate ganglion and supplies the anterior two-thirds of the tongue with taste fibers and the submandibular gland with secretory fibers. The stapedius innervates the stapedius muscle of the ear; the Wrisberg nerve is considered a sensory branch of the facial nerve supplying the tympanic cavity, Eustachian tube and the mastoid area, also the auricle (external ear) and the area in front of it, according to Ramsay Hunt.[10] Thus the facial nerve is to be considered a mixed nerve, carrying not only motor but also sensory and secretory impulses, as stated by Ramsay Hunt, Dejerine,[11] and others.

The muscular branches of the seventh nerve end in the posterior bellies of the digastric and occipitofrontal muscles, muscles of the ear (retrahens aurem and attrahens aurem), the stylohyoid muscle, and all the facial muscles, excluding those innervated by the trigeminus, and including the platysma myoides.

When the facial nerve is affected anywhere in its extent after it leaves its nucleus, a *peripheral* paralysis of this nerve results, known as Bell's palsy.

Etiology.—Lesions of the *pons*, *meninges* (basilar meningitis, tumors), *internal auditory meatus* (tumors of the acoustic nerve), *Fallopian canal*, *mastoid* and parotid regions (accidental or surgical wounds, contusions, pressure—Duchenne, Knapp,[13] Spiller[14]), *suppurations*, so-called *rheumatic* influences, *infectious* diseases (influenza, erysipelas), *tumors* of the posterior skull cavity, such as tumors of the cerebellopontine angle, are some of the causes of a peripheral paralysis of the seventh nerve. In 73 per cent., according to Oppenheim, the cause is rheumatic, while some writers, as Watterman,[12] refer the cause to the lower resistance of the nerve resulting from heredity, constitutional disorders, shock, trauma, syphilis, obstruction of the flow of secretion from the glands surrounding the nerve at its point of exit from the stylomastoid foramen. Bell's palsy may also be bilateral (facial diplegia). This is discussed elsewhere.

Symptoms.—As previously mentioned, the cardinal feature of a peripheral nerve paralysis is total paralysis of the facial muscles supplied by this nerve, including the upper branches which innervate the muscles of the forehead and the orbicularis oculi (which controls closure of the eye). In a peripheral paralysis of the seventh nerve, therefore, the paralyzed side appears smooth. The patient cannot wrinkle the forehead on the affected side and cannot close the eye, the palpebral fissure appearing larger there than on the healthy side. The corner of the mouth droops, the nasolabial fold is obliterated, the tongue deviates toward the diseased side as the result of facial asymmetry. When the patient attempts to laugh, smile, or cry, the facial asymmetry becomes more evident, whistling, talking, and puckering of the lips being defective. There are some other symptoms described, such as Bell's phenomenon, the Negri sign, the Mingazzini sign, excessive lacrimation or abundance of tear secretion, sensory-secretory disturbances of the tongue, hyperacusis, loss of the conjunctival, corneal and of McCarthy reflexes.

Bell's phenomenon consists in rolling the eye-ball upward and outward when an attempt is made to close the paralyzed eye. It is due to the action of the oblique inferior muscle (third nerve). Though observed in normal states, it is especially pronounced in Bell's palsy and scarcely in central facial paralysis.

The Negri sign is apparent when the patient is directed to look up. The eye-balls, or rather the pupils, are not in line, one being higher (on the paralyzed side) than the other.

The Mingazzini sign can be tested only in incomplete facial palsies. The attempt to open the affected eye, under resistance, reveals weakness of the orbicularis oculi.

The McCarthy reflex consists in fascicular contractions of the orbicularis oculi when the region of the eyebrow is tapped.

The so-called excessive secretion of tears (lacrimation, epiphora) is rather an excessive accumulation of tears, caused by deficient action of Horner's and the orbicularis oculi muscles; the tears cannot be directed and moved into the lacrimal canal.

Sensory-secretory disturbances of the tongue are due to the involvement of the chorda tympani. The anterior two-thirds of the tongue shows loss of the sense of taste, especially for bitter and sour.

The secretory disturbances are revealed by dryness of the mouth, because of deficient secretion of saliva. This is usually pronounced in a bilateral facial paralysis.

Hyperacusis, abnormal sensitiveness to noises or refined hearing, may occur as the result of paralysis of the nervus stapedius, an antagonist of the tensor tympani. The tension of the membrani tympani, through the unopposed action of the stapedius muscle, is excessive and causes abnormal hearing. Loss of the corneal, conjunctival, and of McCarthy's reflexes are self-explanatory, since the efferent motor branch, causing these reflexes, is inactive. The mucous membranes are often inflamed on account of defective closure of the eye which, remaining unprotected, is easily subject to irritations.

Contrary to the motor symptoms which are generally very marked, the sensory symptoms in Bell's palsy are unusual. Some cases begin with severe pain in the mastoid region and around the ear; in some cases it radiates to the face and occiput. According to Ramsay Hunt the pain is due to the involvement of the sensory branch of the facial nerve, Wrisberg's nerve, and not to "concomitant lesion of the trifacial or the auricular branch of the vagus" (Webber, 1878). It may be present in mild and severe cases, being of little, if any, prognostic value, that is, the presence of pain does not mean that the case will run an unfavorable course. In some cases hypesthesia or anesthesia was observed on the paralyzed side of the face (Frankl-Hochwart), tongue (Bernhardt), in the concha (Gowers), but such cases are rare.

The electrical reactions are of great importance. In facial paralysis, due to a central lesion, they are always absent, but, as a rule, are present in the peripheral type. The faradic and galvanic excitability of the nerve and muscles supplied by it are usually retained for 10 to 12 days. After this time the galvanic muscular (direct) excitability may become increased. The faradic and galvanic (indirect) excitability may be lost for both the nerve and muscles. This is a complete reaction of degeneration, with flabby, wormlike contractions of the muscles, and the anodal closure contraction stronger than the cathodal (ACC > CCC). There may be an incomplete reaction of degeneration, either nerves or muscles, or both, responding slightly to either current with the galvanic formula reversed. The galvanic formula may, however, be normal, while stimulation with the faradic current will fail. There may also be quantitative changes, stronger currents being required to evoke a neuromuscular action.

Differential Diagnosis.—The diagnosis of a seventh nerve peripheral lesion is not difficult when both sides of the face are compared. The asymmetry becomes evident, especially when the patient tries to perform active voluntary movements of the face, such as wrinkling his forehead, showing his teeth, puckering his lips, or when he is asked to perform some mimical act, such as smiling and laughing. Both mimical and voluntary movements are lost in Bell's palsy, which involves the entire half of the face. In central lesions, for instance, in cerebral hemorrhages, bulbar or pseudobulbar paralysis, only the lower half is paralyzed, with the loss of either mimical or voluntary movements. The paralysis, in other words, may be dissociated. For instance, a person with a cerebral lesion may exhibit no facial paralysis when he shows his teeth, but it will become apparent when he smiles, or laughs. The electrical changes are absent in central lesions, and other signs are usually present indicating central involvement of a nerve.

Course and Prognosis.—The course is more or less acute or subacute. It begins acutely, preceded sometimes by pain, and in children by fever, and headache, according to Oppenheim. A moderate or protracted onset indicates a slow involvement of the nerve, such as the pressure from a tumor; the prognosis in such cases is bad. In favorable or typical, so-called rheumatic types, the course is subacute, lasting from several weeks to several months, usually ending in recovery without any active treatment, for the nerve regenerates slowly. The character of the electrical manifestations may indicate the course and outcome. Absence of electrical changes within the first two weeks is insignificant, but no changes within the third week usually indicate a speedy recovery, within a week or two (mild form). Presence of the reaction of degeneration within the first two weeks means that the course will be protracted, extending over two, three, or more months (severe form). The incomplete reaction of degeneration indicates a course of about two months (middle form). In some cases these rules do not hold. There may be absence of reaction of degeneration, the muscles and nerves responding to both currents, yet remaining paralyzed, as was shown in the case of Babinski and Bouchard.[16] It may also happen that some muscles, as the frontal, will recover, while others will remain spasmodic or flaccid. Such possibilities are likely to occur in traumatic lesions of the facial nerve, when some branches have undergone greater injury than others. But even in the non-traumatic forms, the recovery may be incomplete, resulting in secondary contractures of the affected side, associated movements, and involuntary muscular contractions, so-called facial partial spasms. In the secondary contractures, the affected side shows an increased muscle tonicity, the opposite side, when compared, appearing flaccid or flabby, giving the impression that the healthy side is the paralyzed one. In associated movements, closure of the eye, for instance, is accompanied by raising the corner of the mouth, while, in spasms, muscular twitchings occur in some of the muscles of the affected side. The cause of such undesirable complications has been sought in an abnormal regeneration of the muscle fibers or in shrinkage of the degenerated fibers. Whatever the cause, it cannot be due to electrical treatment, for it occurs in varying degrees in patients who have had no treatment whatever.

Treatment.—It is better to refrain from any active treatment during the first two weeks, during which time mere palliative or symptomatic treatment should be given—heat (dry or moist), pyramidon for relief of pain. After two weeks the faradic or galvanic current may be used, according to the condition of the nerve. If it does not respond to the faradic current, the galvanic should be applied. A large electrode is placed to the neck, sternum, and sacral region, where muscles are few; the smaller electrode is applied to the various branches of the facial nerve at the so-called Erb's points, which indicate the superficial position of the nerves, from which they can be stimulated easily. The entire nerve may be stimulated at its exit from the stylomastoid foramen, by opening or closing the current (the anode or cathode, mostly the anode, as there is usually present the reaction of degeneration); or the current is given for two or three minutes without moving the electrode, or by passing the active electrode slowly along the nerve trunks. Massage of the muscles may also be used. When the paralysis is of too long standing, more than six months, surgical treatment may be given to remedy the asymmetry. The facial nerve is cut, and the peripheral stump is inserted or connected with a cut hypoglossal descending branch or a cut spinal accessory nerve. Anastomosis between the seventh and the cut eleventh or twelfth causes paralysis and atrophy of the tongue, trapezius, or sternocleidomastoid muscles as well as associated movements. For instance, raising the shoulder causes opening of the mouth or its retraction. Therefore, Grant[17] uses for anastomoses the spinal accessory and the descending branch of the nervus hypoglossus. The seventh nerve is cut at the stylomastoid foramen, and the eleventh nerve is divided just before its entrance into the sternocleidomastoid muscle. The central stump of the divided eleventh nerve is stitched to the peripheral branch of the cut seventh, so that the paralyzed seventh nerve gets its impulses from the eleventh nerve. The peripheral stump of the latter is stitched to the ramus descendens hypoglossi, and thus prevented from becoming atrophied.

Bilateral Bell's Palsy (Facial Diplegia, Prosopodiplegia).—Bilateral, total facial paralysis, known as facial diplegia, is uncommon. The causes are the same as in unilateral paralysis: Tumors at the base of the brain, aneurysms, tuberculous and syphilitic lesions, Fallopian canal lesions, rheumatic or infectious causes (diphtheria, influenza, acute poliomyelitis), and finally mechanical injuries, such as the application of forceps (Seeligmüller,[18] Edgeworth[19]), or contusions and fractures of the skull (Hubbell;[20] Oppenheim and Hallez;[21] Ransohoff;[22] Hassin, Levy, and Tupper[23]).

Symptoms.—In facial diplegia of peripheral origin the entire face is paralyzed—for both mimical and voluntary movements. The face is expressionless and masklike; talking is difficult, the patient being unable to pronounce labials, such as b, p or m, and is unable to smile or laugh. The lips droop; the eyes remain open; the tongue may be totally dry because of lack of saliva, and the other symptoms and signs outlined in the previous chapter are present on both sides of the face.

The diagnosis, prognosis, course, and treatment are the same as in unilateral Bell's palsy.

The ninth nerve (nervus glossopharyngeus) contains motor, sensory, secretory, and taste fibers. The motor fibers start principally from the nucleus ambiguus (in the medulla), together with the vagus fibers. The sensory fibers originate in the ganglion nodosum and ganglion petrosum. The secretory fibers supply the parotid; the taste fibers probably originate from, or rather end in, a special (small-cell) nucleus, situated beneath the floor of the fourth ventricle external to the twelfth nucleus. The physiologic rôle of this small-cell nucleus, which anatomically consists of various kinds of cells, is not yet definitely established.

The fibers, forming bundles, leave the medulla near the posterior or dorsal margin of the pons, between the olivary bodies and corpus restiforme. They are always accompanied by the nerve bundles of the vagus and spinal accessory nerves, and it is usually impossible to differentiate between the bundles belonging to the ninth and those belonging to the other two nerves. Piercing the dura, the ninth nerve, with the other two nerves (tenth and eleventh), passes through the jugular foramen, where it is provided with a separate sheath, and after emerging from the base of the brain it becomes an independent, separate nerve. Its motor fibers supply the stylopharyngeal muscle, the posterior pillars of the fauces (together with the vagus), and a portion of the esophagus. The sensory fibers supply the posterior part of the tongue, soft palate, tonsils, upper portion of the pharynx, cavum tympani, and the Eustachian tube. As the principal structures with which this nerve comes in contact are the pons, cerebellum, medulla, dura, and jugular foramen, lesions of these structures may cause paralysis of the ninth nerve.

Tumors of the base of the brain, at the jugular foramen or at the base of the skull, will affect this nerve, but no isolated involvement of it can be expected because of its anatomic relations. The symptoms, in other words, will be mixed or combined with those involving the tenth and eleventh nerves, which will be described in the next chapter. Specific ninth nerve paralysis causes anesthesia of the upper portion of the pharynx, loss of taste in the posterior third of the tongue, soft palate, deglutition difficulties, and loss of the pharyngeal reflex.

The tenth nerve (nervus vagus or nervus pneumogastricus) contains motor, sensory, vasomotor, and secretory fibers. Clinically, the motor portion is the most important, originating with the ninth and eleventh from a common nucleus in the medulla oblongata (nervus ambiguus). The nerve runs, in company with the ninth and eleventh, to the jugular foramen and passes to the neck in the vicinity of the jugular vein and the internal carotid artery. From the neck the nerve enters the thoracic cavity, supplying with motor branches the esophagus, pharynx, larynx, trachea, heart, lungs, and stomach. In addition it supplies the soft palate and pharynx (together with the ninth nerve). Because of the extensive course and the numerous organs supplied by the vagus, the latter gives off a large number of branches, of which the most important clinically are the inferior laryngeal and pharyngeal nerves. In fact the symptomatology of a diseased vagus is practically centered in these two nerves. The superior laryngeal nerve is of much less importance as it supplies mainly the mucosa of the larynx, epiglottis, and the cartilages, while the inferior laryngeal supplies all the intrinsic muscles of the larynx except the cricothyroid, which is innervated by the superior laryngeal nerve.

Etiology.—The causes may be classified as intracranial and extracranial. The intracranial causes are basilar meningitis, tumors of the posterior skull cavity (such as cerebellopontine angle tumors), hemorrhage, and skull trauma. Extracranial causes may be divided into vascular, respiratory, digestive disturbances, infections (Felix[24]), and intoxications. The most common vascular cause is an aortic or subclavian aneurysm, for as the right laryngeal inferior nerve winds around the subclavian artery, it comes in contact with the aortic arch on the left. Pericarditis is another frequent cause, especially in children. *Respiratory* causes of vagus lesion are considerably more important and more common in children than the previous group. Pleurisy, apical tuberculosis, and especially hypertrophy of tuberculous or syphilitic glands situated inferiorly and posteriorly to the larynx may cause compression of the inferior laryngeal nerve. Similar hypertrophy may occur in the glands of the neck, press on the vagus, and cause various vagal symptoms. This pressure etiology (on the neck or within the thorax), so to speak, may be considered the most important and the commonest cause of vagus disorders, especially in children.

Other very important etiologic factors are the infections, of which *diphtheria*, in pediatric practice, plays the most prominent part. In adults the vagus is often affected from tabes, causing laryngeal tabetic crises. In children such etiology is so far unknown.

Symptomatology.—A vagus lesion manifests itself principally in paralysis of the inferior laryngeal and pharyngeal nerves, that is, in paralysis of the soft palate, larynx, and pharynx. In bilateral paralysis the soft palate is totally paralyzed, hangs motionless upon phonation; deglutition, especially for food, is greatly impaired. The vocal cords are paralyzed; the voice is totally lost; the breathing is labored and noisy (inspiratory stridor). Peripheral vagus lesions, however, being usually unilateral, produce much milder symptoms; the soft palate in unilateral vagus paralysis is affected only on one side, and is associated with unilateral paralysis of the larynx, both resulting in a so-called hemiplegia of the soft palate and larynx as noted in Avellis' syndrome.[25] The voice is harsh and hoarse and the respiration is somewhat labored. In tumors of the posterior skull cavity peculiar, so-called vagal attacks occur—changes in pulse (quick or slow), labored respiration, fainting, or flushing of the face, palpitation of the heart. Cushing considers these symptoms the result of medulla involvement (compare chapter on epilepsy).

Prognosis of vagus paralysis is always serious, especially when the cause is an inoperable tumor. In cases of syphilitis and diphtheria, as well as cerebellopontine tumors, a better prognosis may be given.

Course and **treatment** depend on the etiology. Mercury or arsenic preparations are indicated in syphilis. Antitoxin is advocated, probably unjustly, for post-diphtheric paralysis, and surgical treatment for intracranial or neck tumors.

The eleventh nerve (**nervus accessorius Willisii**) is commonly considered as partly a bulbar, partly a spinal nerve. The medullar, upper portion originates in the nucleus ambiguus, together with the vagus and glossopharyngeal nerves, for which reasons the nucleus ambiguus is also called the vago-accessorio-glossopharyngeal nucleus. The spinal portion starts from the motor cells of the upper fifth or sixth spinal cord segments. Obersteiner proposes to consider as the spinal accessory nerve only the portion originating in the spinal cord, referring its bulbar portion to the vagus. The nerve formed by the several spinal roots runs upward into the skull cavity, entering it through the foramen magnum. Joining, in the cranial cavity, the bulbar roots coming from the nucleus ambiguus, the nerve runs to the jugular foramen, where it occupies the most superficial position to the glossopharyngeal and vagus nerves. Leaving the jugular foramen, the nerve runs along the neck between the occipital artery and the internal jugular vein, covered by the sternocleidomastoid and trapezius muscles, which it supplies. The former is supplied exclusively by the eleventh nerve, the latter partly by the eleventh and partly by the upper four cervical nerves. The spinal accessory nerve is exclusively a motor nerve.

Etiology.—As the nerve passes along the vertebral column upward to the foramen magnum, through the meninges to the jugular foramen, along the neck, any morbid conditions in these regions are liable to cause its paralysis. Thus, spondylitis and meningitis (tuberculous, syphilitic), tumors of the spine and the meninges, lesions of the foramen magnum (tuberculosis of the base or its fractures), lesions of the jugular foramen (tumors, enlargement of neck glands from syphilis, tuberculosis), foreign bodies or injuries (wounds, accidental or surgical), are all likely to produce a paralysis of the eleventh nerve.

Symptoms.—Unilateral lesions cause paralysis of the sternocleidomastoid and a partial paralysis of the trapezius muscles on the corresponding side. An abnormal position of the head will result, the head being turned toward the healthy side, with rotation of the chin toward the diseased shoulder. In bilateral paralysis, the head will fall forward. Paralysis of the trapezius muscle is generally less pronounced. When present, the shoulder cannot be raised. It droops, and turning the head backward is impaired. If the lesion is in, or near, the jugular foramen, the foregoing symptoms are accompanied by those of paralysis of the tenth nerve, such as unilateral paralysis of the soft palate and larynx, and other features previously outlined. The unilateral involvement of both the tenth and eleventh nerves produces what is known as Schmidt's syndrome, namely, unilateral paralysis of the soft palate, larynx, the sternocleidomastoid and trapezius muscles.

Prognosis, course, and treatment are the same as outlined for the tenth nerve.

The twelfth nerve (nervus hypoglossus) arises from a powerful nucleus in the fourth ventricle. The fibers leave the medulla between the pyramids and the olivary bodies, pierce the dura, and leave the posterior skull cavity through the anterior condyloid foramen. It supplies the intrinsic and extrinsic muscles of the tongue. The function of this nerve is purely *motor*, moving the tongue, otherwise acting as any other motor nerve.

Etiology.—Peripheral lesions of this nerve are comparatively rare, but may occur from disease processes in the posterior skull cavity, such as cerebellopontile tumors, meningitis, basal skull fractures, syphilis, and tuberculosis of the base of the cranium.

Symptoms.—Unilateral paralysis of the hypoglossal nerve affects one-half of the tongue, which appears atrophied (Fig. 49) and corrugated, showing fibrillary twitchings. There is some difficulty in talking, eating, and moving the tongue, which, when protruded, is deviated toward the diseased side, with the tip turned to the healthy side. Especially pronounced are some of these symptoms when the hypoglossal nerve is paralyzed on both sides, as in bulbar paralysis. While traveling on the neck from the condyloid foramen to the tongue, it may be involved, in the region of the jugular foramen, with the tenth or eleventh, or both, resulting in the characteristic syndromes of Jackson, Tapia, and Collet.

In Jackson's syndrome[26] the paralysis of the twelfth nerve is associated with that of the tenth. The symptoms in Jackson's case were right lingual hemi-atrophy and paralysis, combined with that of the right half of the soft palate and the right vocal cord, that is, a combined involvement of the tenth and twelfth nerves. In Tapia's syndrome, the symptoms are the same as in Jackson's, except that, instead of the soft palate, the laryngeal muscles are involved, that is, the tongue and larynx are involved unilaterally. It is, according to Dejerine, an exquisitely peripheral paralysis, like the Jackson and Schmidt syndromes, while Avellis' syndrome (unilateral paralysis of larynx and soft palate), he considers a central bulbar lesion.

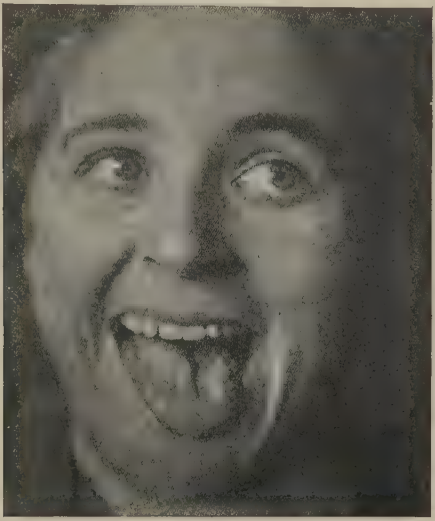

FIG. 49.—The left half of the tongue is greatly atrophied, wrinkled, and covered with furrows. The tongue is deviated to the diseased side.

The last syndrome, that of Collet, is the most instructive example of a combined cerebral nerve involvement, under which name it was described by Joseph Beck[28] and myself, independently of Collet.[29] As this syndrome is of great clinical interest, it deserves a more detailed discussion, which follows.

Combined peripheral cerebral nerve involvement (ninth, tenth, eleventh and twelfth) or syndrome Collet, and syndrome of the jugular foramen (Vernet). Peripheral unilateral involvement of several cranial nerves is quite rare, as compared with a central lesion caused by brain lesions proper, or by basilar meningitis. In the jugular foramen syndrome (Vernet), the ninth, tenth, and eleventh nerves are involved, as they are all grouped together in this foramen, and after leaving it they are enveloped by a common sheath. Around and near the foramen are situated groups of lymph-nodes (the deep neck glands of Krause). Enlargement of these glands, as was the case in my patient, or their hypertrophy from syphilis, tumors of the base of the skull, foreign bodies (bullets, for instance), may cause involvement of all these three nerves alone or including the twelfth nerve. Vernet collected and reported 26 cases of the former (three nerves), including his own cases, while but two instances of lesions of the four nerves were recorded, one by Collet and the other by Dr. J. Beck and myself. In Collet's case the lesion was caused by a bullet which lodged behind the mastoid process. The patient complained of difficulty in swallowing, especially hard food, and buzzing and deafness in the right ear. The tongue was slightly deviated to the right, the right vocal cord and the right half of the velum and soft palate were paralyzed, and the right shoulder drooped. During phonation the entire right half of the posterior pharyngeal wall was inactive and drawn to the left by the overaction of the healthy side; taste for bitter, sweet, and salt were lost on the right. The findings are summed up by Collet as hemiplegia of the tongue (twelfth) and of the trapezius muscle (eleventh). In addition the pulse was accelerated (tenth nerve) and pain radiated from the angle of the lower jaw to the shoulder. The simultaneous peripheral involvement of the last four cranial nerves which took place in this case may result, according to Collet, from a lesion in front of the occipital condyle not more than 1 cm. in diameter, yet sufficient to injure all the nerves passing the jugular and anterior condyloid foramina.

Analogous to this case was the one recorded by J. Beck and myself. The patient was an 18 year old girl who had complained for two years of continuous pain in the left side of the neck, face, and forehead, of difficulties in deglutition, and of speech troubles. At the age of six she underwent an operation for tuberculosis of the cervical glands. At the age of 13 torticollis developed on the right. At the age of 16 speech impairment was noticed; the voice became rather hoarse and nasal, deglutition became more difficult, especially with solid food.

The examination revealed very indistinct nasal dysarthric speech; a slightly hoarse voice, interrupted by stridor-like breathing; atrophy of the left sternocleidomastoid and the upper portion of the trapezius muscles; paralysis of the left half of the soft palate, uvula, and the left vocal cord; atrophy of the left half of the tongue (Fig. 49), with marked fibrillary twitchings, and deviation to the left when it was protruded; rotatory nystagmus, especially upon looking to the extreme right or left; a bilateral otosclerosis and deafness. The pharyngeal reflex was greatly diminished. The pulse was 144.

The principal findings, aside from the nystagmus and deafness, were confined to the ninth, tenth, eleventh, and twelfth nerves, which were jointly affected in the neighborhood of the left jugular foramen, probably by a calcified tuberculous gland, in view of the history of tuberculosis of the cervical glands. A Roentgen-ray examination revealed a calcified mass in that neighborhood or region, which was dissected by Dr. Joseph Beck. Remarkable improvement occurred after the operation, and upon examination, six months afterward, scarcely any atrophy was present; but the fibrillary twitchings did not disappear. The soft palate, uvula, and deglutition troubles and difficulties in breathing and of speech had disappeared. There were no changes in the reflexes, nor in the nystagmus, deafness or otosclerosis. The nystagmus and otosclerosis had, of course, nothing to do with the nerve involvement. About two months after the last examination, or eight months after the operation, the symptoms returned, and a second operation revealed that the previously mentioned nerves were surrounded by a dense fibrous tissue. Though marked improvement occurred after the second operation, also it was followed by exacerbations of the symptoms, to which the patient finally succumbed. There is no doubt that cases of combined extracranial nerve involvement are of rare occurrence, yet it is imperative to recognize them early, when they are amenable to surgical or other treatment, which, as in any other peripheral nerve lesion, depends on the etiology.

(C) PALSIES OF SPINAL NERVES

Contrary to the cranial nerves which usually run to the periphery singly, except the ninth, tenth, and eleventh, the spinal nerves ordinarily form combinations known as plexuses. Of the five plexuses—cervical, brachial, lumbar, sacral, and coccygeal—the first four are of great clinical value and will be considered at some length.

Cervical Plexus Paralysis.—The cervical plexus is formed by the anterior divisions of the upper four cervical nerves, which form three loops. The fourth cervical nerve is connected with the brachial plexus. The cervical plexus is situated on the neck, above the median scalene muscle, covered by the upper portion of the sternocleidomastoid. This plexus supplies numerous nerves to the skin and muscles of the neck.

Clinically the most important is the phrenic nerve which, derived from the fourth cervical, supplies the diaphragm, the pleura, and the pericardium. The right phrenic nerve comes in contact with the right innominate vein, the left with the aortic arch.

Etiology.—The causes of unilateral, or bilateral paralysis of the **phrenic** nerve are infections (diphtheria, pneumonia, typhoid fever), intoxications (alcohol, lead), tumors of the mediastinum, vertebral column lesions (tuberculosis, syphilis, tumors), meningeal involvement of the upper cervical region.

Symptoms.—The result of a phrenic nerve lesion is paralysis of the diaphragm, an important respiratory muscle. There is difficulty in breathing, which is carried on by the upper portion of the chest, the lower half remaining inactive. The epigastric region during inspiration does not protrude, but retracts. As the diaphragm is higher than normal, there is an elevation of the liver which is inaccessible to palpation or percussion. Active breathing, as forced, deep inspiration, or coughing and expectoration, are impossible, and this may cause deficient ventilation of the lungs and lead easily to pneumonia.

The **prognosis** is serious, especially in bilateral phrenic lesions, or when, as in pneumonia, excessive breathing is necessary.

The **course** and **treatment** depend on the fundamental disease. Severe attacks of dyspnea can be stopped by administering morphine. Otherwise absolute rest, as in treatment of acute myocarditis or endocarditis, should be insisted upon.

Brachial Plexus Palsies.—The brachial plexus is formed by the fifth, sixth, seventh, and eighth cervical nerves, and the first thoracic or dorsal nerve. Soon after leaving the intervertebral foramina, these nerves combine, forming groups termed trunks and cords. The fifth and sixth cervical nerves form the upper trunk; the eighth cervical and first dorsal, the lower trunk, the middle trunk being formed by a single *seventh* cervical nerve. These so-called primary trunks divide into three anterior and three posterior divisions, the latter forming the secondary *posterior* trunk, or posterior cord; the *anterior* division of the upper and middle trunks form the *outer secondary* trunk, or cord, while the remaining anterior division of the lower trunk independently forms the *inner* cord, or *secondary inner trunk*. From the resulting posterior, outer, and inner cords, terminal branches arise to be distributed to the muscles of the scapula, shoulder, and upper limb.

The brachial plexus occupies an area above and below the clavicle, the supraclavicular region being the site of the primary trunks and their branches, the subclavicular region that of the secondary trunks, or cords, where they give off the terminal nerves (circumflex, median, ulnar, etc.). In addition to the latter, before they form cords, the primary trunks and the spinal nerves themselves give off so-called collateral branches, the most important of which are the long thoracic (for the serratus anticus muscle) and the suprascapular nerves (for the supraspinatus and infraspinatus muscles).

The most important structures with which the various components of the brachial plexus come in contact are the vertebræ and their processes, the clavicle, shoulder bones, the subclavian and axillary arteries. The latter are surrounded by the three cords (posterior, outer and inner) their names outer, inner, posterior indicating their positions relative to this artery. Lesions of the foregoing anatomic structures (vertebræ, the shoulder joint, clavicle, arteries), such as aneurysms, fractures, and dislocations, tumors, accidental or surgical injuries to the shoulder joints, are the causes of total or partial paralysis of the brachial plexus.

Total Brachial Plexus Paralysis.—In total brachial plexus involvement the entire upper limb (hand, forearm, upper arm, shoulder) is paralyzed—the paralysis having the characteristics of a peripheral nerve lesion. It is *flaccid*, the entire limb hangs motionless, showing no resistance upon passive movements; the muscles are hypotonic; the reflexes, as the biceps, triceps, ulnar, radial, and others, are abolished; atrophy is widespread, combined with degenerative, electrical changes, and vasomotor and trophic disturbances. Sensory symptoms are often present, such as anesthesia of the hand, forearm, and postero-external surface of the upper arm, while the inner aspect of the arm is always intact, being innervated by the intercostal nerves. Oculopupillary symptoms (narrowing of the palpebral fissure and of the pupil, enophthalmus, flattening of the cheek) may occur on the affected side. Vasomotor and trophic disturbances, such as cyanosis, edema, and glossy skin, are frequent.

In children a total brachial plexus paralysis is one of the varieties of obstetric paralysis which will be considered in one of the next chapters.

The **diagnosis** is quite easy, provided sensory symptoms and oculopupillary manifestations are present, in connection with a reliable history. In the absence of such history or the absence of sensory and oculopupillary symptoms, it is almost impossible to differentiate it from poliomyelitis.

Prognosis.—As to life this is good, but as to permanent recovery, especially in children, it is always serious, for wasting of the muscles may be complicated by atrophy of the bones of the affected extremity, and become permanent.

The **course** is protracted and is often associated with severe pain.

Treatment depends on the etiology. Usually there has been some trauma, which calls for surgical measures.

Partial Brachial Plexus Palsies.—These, comparatively, occur much more frequently in the form of upper (Duchenne-Erb type) or lower (Klumpke-Dejerine type) brachial plexus paralysis.

The Duchenne-Erb type is the usual variety of partial brachial plexus paralysis. Nerves are involved which are formed principally by the fifth and sixth cervical nerves, namely the nervus musculocutaneous (which supplies the biceps, coracobrachialis, and brachialis anticus muscles), musculospiral nerve (the branch supplying the supinator longus muscle), circumflex nerve (musculus deltoideus) and nervus suprascapularis (for the supraspinatus and infraspinatus muscles). All these nerves may be simultaneously set in action by stimulation at the so-called Erb's point. This is situated in the supraclavicular fossa, 2 or 3 cm. above the clavicle (about two fingerbreadths) immediately outside the posterior border of the sternocleidomastoid, at the level of the anterior tubercle of the transverse process of the sixth cervical vertebra. Stimulation at this point results in a contraction of the deltoid, biceps, brachialis anticus, and supinator longus muscles. In paralysis of these muscles it is impossible to flex and supinate the forearm (nervus musculocutaneous) or to abduct and rotate the arm (circumflex nerve); the affected limb appears extended, inwardly rotated, pronated, and adducted. Movements of the hands and fingers are possible. In some cases only the deltoid muscle, that is, only a part of the brachial plexus, may be involved.

Sensory changes may be either absent, or mild. If present they are confined to the extensor aspect of the arm and forearm. Electrical and trophic changes (muscular atrophies) may be present as in any other peripheral nerve lesion.

Paralysis of the Klumpke-Dejerine type, or lower brachial plexus paralysis, involves the lower primary trunk and its nerves, the eighth cervical, and the first dorsal. The main branches emanating from this trunk are the ulnar nerve and a portion of the median, therefore, the symptoms of Klumpke paralysis will be atrophy of the muscles that these nerves supply—the small muscles of the hand (interossei, lumbricales, thenar and hypothenar muscles), resembling progressive muscular atrophy, Duchenne-Aran type. The atrophic paralysis is usually accompanied by sensory disturbances—anesthesia of the inner (and sometimes the external) aspect of the hand, forearm, and shoulder. Oculopupillary symptoms may be present if the lesion of the plexus is near the cord, where the sympathetic nerve may be involved at the level of the roots. If the ramus communicans of the first dorsal nerve escapes the lesion and only the spinal nerves are involved, the oculopupillary symptoms will be lacking.

This paralysis usually forms a part of the clinical picture of a total brachial plexus lesion, cervical rib, and obstetric paralysis.

Aside from these *three* types of brachial plexus lesion, many authors recognize and describe various *subtypes*, depending on the involvement of the principal spinal nerves before they form primary or secondary trunks. In other words, the palsies are classified according to their localization, some, as Grenet,[32] accepting a radicular type, plexus type, and *terminal nerve* lesion type. The radicular type he subdivides into intradural and extradural, which he also differentiates according to the separate involvement of the motor and sensory roots of the collateral branches and the presence of the oculopupillary symptoms. The latter, for instance, speak for an intradural lesion, while the paralysis of the supraspinatus and infraspinatus muscles, supplied by the suprascapular nerve, indicates an extradural type of the Duchenne-Erb paralysis. Further details of this interesting, though highly artificial, classification cannot be considered here. I wish merely to point out that the *roots* of the brachial plexus are not the spinal cord roots but the spinal nerves (fifth, sixth, etc.) before they form a plexus and give off ramifications, a fact pointed out by Klumpke.[31]

Obstetric or Birth Palsies.—As the name indicates, some paralyses in children may be obstetric in origin. They are usually the result of undue traction on the shoulders in a breech extraction or may follow difficult forceps deliveries. Brain, spinal cord and peripheral nerve injuries may result. The two varieties, caused by cerebral or spinal cord injury, cannot be discussed here, this chapter being devoted only to peripheral nerve injuries. These injuries constitute the most common type of obstetric palsies, affecting, usually unilaterally, either the facial nerve or the brachial plexus. Ordinarily, a portion of the latter is involved, such as the upper, the entire brachial plexus rarely being affected. In other words, birth palsies occur usually in the form of Duchenne-Erb paralysis. They are caused by injuries which an obstetrician may inflict upon the child during manual or instrumental delivery. For instance, hooking the fingers in the arm-pit to extract the infant, or traction by forceps may cause, according to some authors, a direct pressure on, or tearing, overstretching or rupture of the nerves. Forcible depression of the shoulders with the head bent to the opposite side (Kennedy[33]), asphyxia (Stransky[34]), misapplied forceps slipped over the cervical region, the head being too small and not flexed (Fieux[35]), hemorrhages in the plexus (Clark, Prout and Taylor[36]) were all looked upon as the probable causes of injury and paralysis of the brachial plexus. Lange[37] believes that distortion of the shoulder joint capsule, during labor, is responsible. The same etiology is advocated by Thomas,[38] who considers the brachial birth palsy secondary to the joint involvement (spraining, tearing, dislocation, etc.), insisting that the shoulder joint should be examined for possible trauma in every newborn infant.

The **differential diagnosis** is not difficult if the etiology is certain, otherwise, as stated previously, it may be impossible to distinguish it from anterior poliomyelitis. Sever[39] points out that atrophy in obstetric paralysis is never very marked, except in some cases in which the lower arm is affected, and it is never so extreme and noticeable as in cases of infantile paralysis.

Prognosis depends upon the severity of the lesion, its extent, and treatment. In mild cases in which only the nerves are compressed, no hemorrhages occur, and electrical changes are absent, the prognosis is good, and cure can be obtained from the faradic or galvanic current, as Duchenne pointed out. In severe cases in which hemorrhages are present, the nerves torn and lacerated, and the proper treatment (surgery) has not been employed, the prognosis is bad. The muscles and bones of the affected limb become permanently atrophied; there is arrest of development of the bones, according to Huet;[40] the shoulder and elbow joints become loose, as Bullard[41] notes; the affected limb may become shorter, as reported by D'Astros[42] and Sever. Paralytic deformities are more likely to occur in this condition than in plexus paralysis of any other origin.

At any rate, as Warrington and Jones[43] point out, in birth palsies of any severity, a less favorable prognosis can be given than in plexus paralyses in adults. Hence, it can easily be understood how important it is to recognize these conditions early, and urge the employment of surgical measures in suitable cases.

Treatment.—Prophylactic treatment—avoiding unnecessary manipulations especially by unskilled hands—is most important. (Guillemot[44] reports 30 cases of brachial plexus birth palsies which were caused by an unskilled midwife.) If the paralysis is mild, rest, proper position and support of the limb, electricity, and massage may lead to recovery (complete or partial). Kennedy waits two months, Taylor[45] longer, about a year, unless the electrical examination shows far advanced changes. Thomas recommends deferring operations on the brachial plexus, its roots or branches until a paralysis from inclusion of the nerves in cicatricial tissue is eliminated. Sever and Boorstein[46] advise orthopedic treatment: a plaster cast embracing the thorax and the paralyzed limb. In the articles by Kennedy, Taylor, and Sever, the operative technique and results are reported in detail.

Paralyses of the terminal branches of the brachial plexus occur in children much less frequently than in adults, and they will be considered here only in general.

Paralysis of the long thoracic nerve (posterior thoracic nerve, external respiratory nerve of Bell). This nerve originates from the fifth, sixth, and seventh cervical nerves before the latter form the primary trunks, and supplies the serratus magnus muscle. The latter arises from the side of the thorax (from the first to the ninth or tenth rib) and is inserted in the vertebral border of the scapula. It fixes the scapula firmly to the thorax, helping the trapezius to raise the shoulder and the arm vertically.

Symptoms.—Paralysis of the long thoracic nerve involves the serratus magnus muscle, causing a so-called "winged" scapula, so that the hand can be passed between it and the thorax. The arm cannot be raised to the vertical position. Sensory disturbances are absent.

Causes.—These are the same as in paralysis of the brachial plexus, namely trauma, and infections (diphtheria, typhoid, influenza).

Prognosis is good, especially in the infectious type, and the treatment depends on the etiology (electricity, surgical measures).

Nervus Suprascapularis.—This nerve arises from the fifth and sixth cervical nerves, just at the point at which they are ready to form the upper primary trunk. Covered by the trapezius muscle, it runs to the suprascapular notch, dividing, after crossing it, into two branches which supply the supraspinatus and infraspinatus muscles. The former, together with the deltoid, abducts the arm; the infraspinatus muscle externally rotates and adducts the arm when it is elevated.

Symptoms.—Isolated paralysis of the suprascapular nerve is very uncommon. It will result in paralysis of the supraspinatus and infraspinatus muscles, causing a shoulder droop; the arm can be raised only to the horizontal position. Attempts, for instance, to comb the hair will be inhibited in paralysis of the infraspinatus.

Causes, Prognosis, Course, and Treatment.—These are the same as in every other peripheral nerve lesion.

Circumflex Nerve (Nervus Axillaris).—This nerve is a branch of the posterior cord of the brachial plexus, being derived from the fifth and sixth cervical nerves. It runs through the axilla and clavicular fossa, and supplies the deltoid muscle and teres minor. The function of the deltoid is to raise the arm and abduct it, the teres minor rotates the humerus.

The **symptoms** of a circumflex nerve paralysis are, therefore, paralysis of these two muscles. The arm cannot be raised or abducted. The patient is unable to dress himself, to carry a spoon to his mouth, etc. The impairment of function of the deltoid will be partial, as the neighboring muscles (supraspinatus, infraspinatus, trapezius, may take up its functions. Sensory disturbances (anesthesia) may be present in a small patch over the deltoid muscle. The diagnosis in long-standing paralysis can be made easily from the atrophy of the deltoid, from the absence of contours of this muscle, and from the lax condition of the humerus.

Causes.—These comprise trauma, pressure, and intoxications (lead).

Nervus Musculocutaneus.—This originates from the *outer* cord of the brachial plexus, the fifth, sixth, and seventh cervical nerves. It supplies the flexors of the forearm—biceps, coracobrachialis and brachialis anticus. The sensory fibers (nervus cutaneus lateralis) supply the outer, anterior, and posterior surfaces of the forearm as far as the wrist (posterior terminal branch) and, through the anterior terminal branch, the middle portion of the thenar eminence. The sensory fibers form anastomoses with the radial nerve. Symptoms of a traumatic or other paralysis of this nerve are a flaccid atrophic paralysis of the flexors of the forearm. The latter remains extended, though flexion may take place from the action of the supinator longus. Sensory disturbances, if present, are confined to the outer surface of the forearm.

Musculospiral (Radial) Paralysis.—This is the largest nerve given off by the brachial plexus, originating, like the circumflex nerve, from the posterior cord (fifth, sixth, seventh, and eighth cervical nerve roots). Crossing the axilla it runs along the posterior surface of the upper arm, accompanying the superior deep artery into the interval between the inner and outer heads of the triceps muscle. Winding around the spiral groove of the humerus, the nerve appears on the external and afterward on the antero-external surface of this bone. Above the bend of the elbow, it terminates by dividing into the radial and posterior interosseous branches. Before it divides into the terminal branches, the nerve gives off two cutaneous branches—the internal cutaneous nerve, for the postero-inferior surface of the arm down to the elbow, and the external cutaneous branch, for the postero-external aspect of the arm and posteromedian surface of the forearm as low as the wrist, anastomosing with the cutaneous branch of the musculocutaneous nerve (Morris).

The *muscular* branches supply the triceps, anconeus, supinator longus and supinator radii brevis, extensors carpi radialis longus and radialis brevis, extensor and abductor pollicis, extensor indicis, extensor digitorum communis, extensor minimi digiti and extensor carpi ulnaris. In short, the musculospiral nerve supplies the supina-

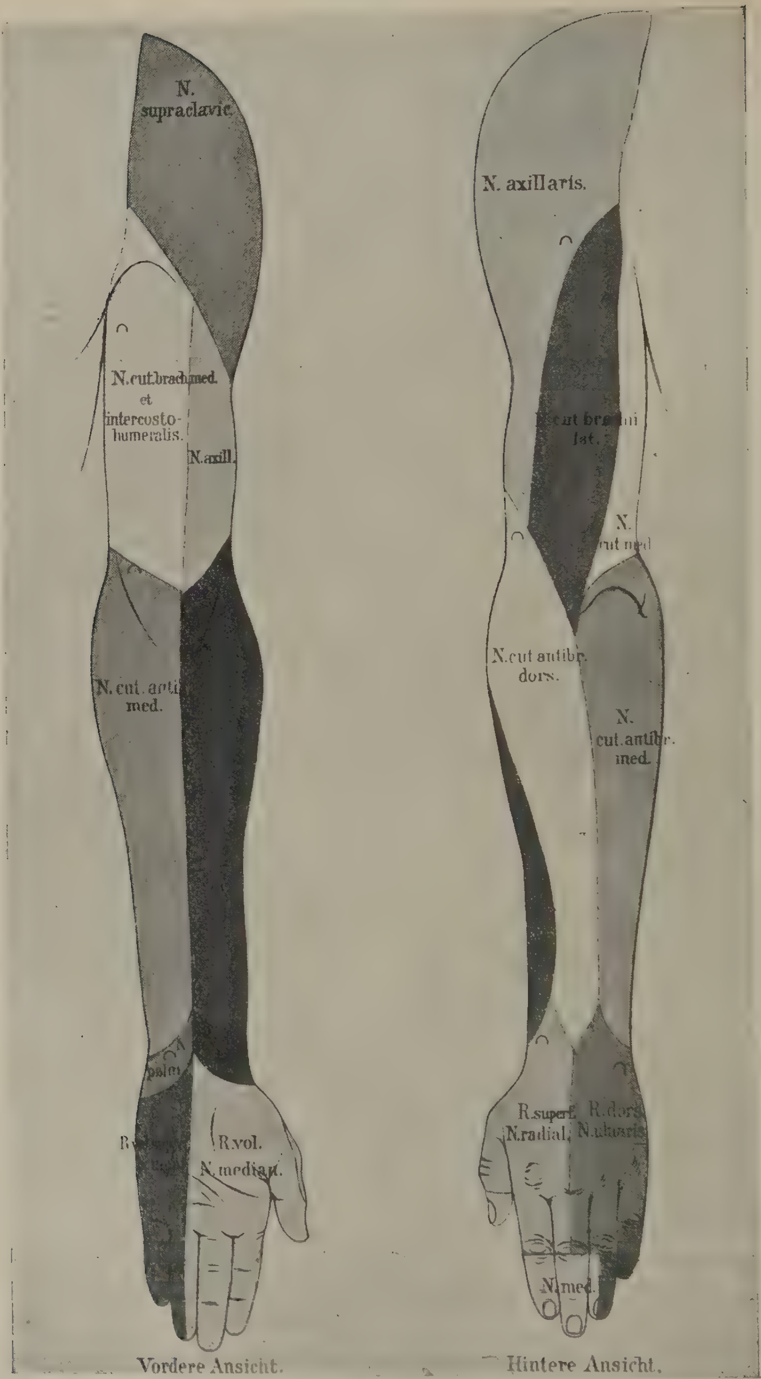

FIG. 50.—Sensory areas of the upper extremity (from Hasse's Atlas).

tors and extensors of the forearm, wrist, thumb, basal phalanges of the fingers and the abductors of the thumb and wrist.

The sensory portion (called also radial nerve) supplies the radial surface of the dorsum of the hand, the dorsal surface of the thumb, of the index finger and the radial half of the middle finger, except the terminal phalanges, which are supplied by the median nerve (Fig. 50).

Symptoms.—Lesion of the musculospiral nerve may cause paralysis of all the foregoing muscles if situated high enough, that is, when the paralysis is total. This occurs in dislocations or fractures of the humerus, and in crutch or other pressure palsies. More frequently, the paralysis is partial, involving only some of the muscles innervated by the musculospiral nerve, depending on the seat of the lesion. The usual seat is the forearm, above or below the origin of the supinator longus muscle. In the latter the triceps and anconeus will be spared; the olecranon or triceps reflex will be retained, but the rest of the extensors, as well as the flexor of the forearm (musculus supinator longus) will be paralyzed. Flexion of the arm will be accomplished by the biceps; the contraction of the belly of the supinator longus will be lacking. This lack of supinator longus contraction is, according to Tinel,[47] the most characteristic, pathognomonic sign of musculospiral nerve paralysis. If the latter is involved below the origin of the supinator longus, the latter will be spared, and even the wrist extensors will be spared, so that the lower the seat of the lesion, the fewer the muscles that are involved. On the other hand, in such cases sensory disturbances are likely to be *more* pronounced. For instance, lesion of the musculospiral nerve close to the wrist may cause anesthesia of the radial side of the thenar eminence and the thumb, and of the hand and forearm if the antibrachium cutaneus lateralis is also involved (Kramer). The most *characteristic* symptom of musculospiral paralysis, if the lesion is not too low, is wrist-drop, combined with inability to abduct and extend the thumb and supinate the forearm. In higher seated lesions, as noted, there will be absence of supinator longus contraction and weakness of the flexors, which is mainly due to the lack of coöperation on the part of the paralyzed antagonists.

In many cases, when a stronger flexion of the fingers can be obtained, the paralyzed extensors contract, causing an extension of the wrist. Extension of the hand may also be accomplished through the action of the pronator teres, especially when the forearm is supinated. These facts show that one should use caution in studying the condition of a paralyzed nerve, keeping in mind intervening nerves and muscles and other factors which cannot be considered here in detail.*

Sensory disturbances, if present, show in the radial half of the dorsum of the hand, the dorsum of the basal phalanges of the thumb, index finger, and the radial half of the dorsum of the basal phalanx of the middle finger (Fig. 50). This is the classical or ideal distribution. In reality it occupies a much smaller area, and is mostly hypesthetic.

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* There are a number of contributions on this subject. Of American contributions the articles of Pollock, Woods and Ingham, all published in the Archives Neurol. and Psychiat., vols. ii and iii, are recommended.

Incomplete lesions of the nerve as tearing, lacerations, prolonged pressure usually cause very little discomfort, such as pain, contrary to what is usually observed in similar conditions in other nerves. Therefore, this nerve does not give the so-called "irritation syndrome" of Tinel,[47] but, according to this author, trophic disturbances are quite common and may be pronounced. Here I would mention adhesions with deformities of the hand, retraction and infiltration of the skin, desquamation, enlargement of digital articulations, the nails being usually spared.

Course and **prognosis** depend on the cause. Electrical examination may give an indication of the condition of the nerve.

The **treatment** may be electrical or surgical. The musculospiral nerve regenerates sooner than any other nerve after an end-to-end suture.

Median Nerve (Nervus Medianus).—The median nerve is formed by fibers derived from the lower three cervical nerves (sixth, seventh, and eighth) and first dorsal, that is, from the outer and inner cords. Descending into the axillary fossa, this nerve runs along the inner aspect of the arm, partly covered by the biceps muscles, and passes between the two heads of the pronator teres muscle. On the forearm it is situated between the superficial and deep flexors of the fingers, and at the lower border of the anterior annular ligament, it breaks up into terminal branches.

It does not supply any muscles on the upper arm; but, together with the ulnar nerve, it supplies all the flexors of the hand and fingers as well as the pronators. Thus, it innervates the pronator teres, pronator quadratus, flexor carpi radialis, palmaris longus, flexor digitorum communis sublimis, the external or radial half of the flexor digitorum communis profundus, flexor pollicis longus, abductor pollicis brevis, opponens pollicis, the superficial head of the flexor pollicis brevis, and the first two lumbricales (on the radial side of the forearm). The sensory branches supply the thenar, central part of the volar surface of the hand, the palmar surface of the thumb, index, middle, and radial aspect of the ring finger, the dorsal surfaces of the terminal phalanges of the middle fingers and thumb as well as the matrix of the nail (Fig. 50).

Symptoms.—According to the location of the injury, the symptoms will be complete or partial paralysis. In the former, when the lesion is on the upper arm, all the muscles supplied by this nerve will be paralyzed. Thus pronation of the forearm and hand will be impossible; the flexion will be preserved only in the fingers supplied by the ulnar nerve (the last three fingers); the hand will be adducted and slightly flexed in the ulnar half, the thumb adducted and extended (ape hand); flexion of the terminal phalanges of the middle and index fingers and thumb will be impossible. With a lesion of the nerve above, or near, the wrist joint, only the small muscles of the hand will be paralyzed, resulting in paralysis of the thenar eminence with the thumb adducted, the hand supinated, and opposition of the thumb and flexion of the terminal phalanges lacking.

The paralyzed muscles, if the paralysis is of long standing, become atrophied, the atrophy being especially evident in the thenar eminence and the radial portion of the lumbricales. Often the atrophy is absent because of anastomoses between the ulnar and the median nerves.

As in musculospiral paralysis, some functions of the paralyzed median nerve muscles may be taken over, or supplemented by other muscles. Thus, Woods points out that in some cases of median nerve lesion pronation was partly possible through the action of the extensors of the wrist and fingers.

Sensory disturbances (anesthesia) may involve the hand (the volar surface to the fourth metacarpal bone), the first three fingers, including the radial side of the fourth finger (their volar aspect) and the dorsal surface of the second and third phalanges of the index and middle fingers. As in radial nerve lesions, the sensory distribution of anesthesias from a median nerve lesion differ individually, and do not correspond to the anatomic distribution of the sensory branches. Vasomotor disorders are evident in cyanosis of the hand and infiltration of the subcutaneous tissue; trophic, as impairment of nail growth.

Causes of median nerve lesions in children are injuries to the nerve itself (wounds—stabbing or gunshot—fractures of forearm and upper arm, dislocations of the shoulder, elbow or wrist joints), in short, any lesion of the structures with which the nerve comes in contact in its course from the brachial plexus to the periphery. Over-exertion, as in piano playing and sewing, also causes a median nerve lesion. Incomplete lesions of the median nerve, caused, for instance, by laceration, compression and stabbing of the nerve, will give signs and symptoms of a *neuritis*. There is usually severe pain, spontaneous and increased from pressure, or active or passive movements, hyperesthesias, hypesthesias and anesthesias, and marked trophic disturbances. The motor phenomena may be less pronounced, the *pain* and the *trophic* disturbances being the predominating features. The latter appear as disorders of the nail growth in the thumb, index, and middle fingers. According to Tinel, trophic disturbances in the *nails* are "absolutely constant" in median neuritis, being, in mild cases, its sole manifestation. Fibrosis of the tendons of the flexor muscles, causing irreducible immobilization of the corresponding fingers and thumb, and retraction of the palmar fascia, adhesions of the skin to the fingers, fibrosis of the interphalangeal joints, are other types of trophic lesions in median neuritis (Tinel). The pain is, in some cases, the predominating symptom, often occurring in the form of so-called *causalgia*. The pain is of burning character, persisting day and night, radiating to the shoulder and increasing on the slightest touch, or psychic excitement. There is no paralysis, no anesthesia, and but slight vasomotor or trophic disturbances. Hot poultices are well borne, being the only means of relieving the severe pain. The cause is probably a lesion of the sympathetic nerve fibers. The **course** is subacute or chronic. Tinel says it may last 15 months, and gradually subside.

Treatment of Causalgia.—Alcohol injections or nerve resection proved useful in some cases.

Ulnar Nerve.—The ulnar nerve, derived from the outer cord (eighth cervical and first dorsal nerves), runs to the forearm between the internal condyle of the humerus and the olecranon process of the ulna, passes between the two heads of the flexor carpi ulnaris, reaching the wrist under cover of this muscle. Above the latter it pierces the deep fascia, crosses the annular ligament near the pisiform bone, terminating in superficial and deep divisions.

This nerve, together with the median, supplies the flexor muscles of the forearm, hand, and fingers. On the forearm it supplies the flexor carpi ulnaris and the inner portion of the flexor digitorum profundus; on the hand it supplies all the muscles of the hypothenar (abductor minimi digiti, flexor brevis, and opponens minimi digiti) two inner lumbricales, all the interossei, the adductor of the thumb, and the inner or deep head of, the flexor pollicis brevis. The sensory branches supply the hypothenar and the little finger, as well as part of the ring finger (Fig. 50).

Symptoms.—The foregoing muscles are all involved when the ulnar nerve is totally paralyzed, causing a complete paralysis of the wrist and last two or three fingers. The result will be lack of, or insufficient, flexion of the hand, only the radial or external flexion being possible. The flexion of the hand will be accompanied by radial abduction. Paralysis of the inner lumbricales will cause a paralysis of the ground or basal phalanges—they will remain extended (from the action of the extensor digitorum communis), while the middle and terminal phalanges will be flexed (from the action of the flexor digitorum communis longus). Such a position of the fingers—extended basal phalanges with the terminal ones flexed—is called claw hand or main en griffe. Claw hand may not be manifest if the flexor digitorum profundus is simultaneously paralyzed, or it may be simulated by adhesions or contractures. Paralysis of the thumb (adduction and its opposition) is another characteristic feature of ulnar paralysis, as well as lack of adduction and abduction of fingers (interossei muscle paralysis).

Paralysis of the ulnar nerve may be masked by supplemental movements. For instance, the abduction of the fingers may be accomplished by the extensor communis digitorum, i.e., by the musculospiral nerve, but then the abduction will be accompanied by their extension. On the other hand, the flexors of the fingers are also their adductors, and the patient will be able to adduct or approximate the fingers by flexing them, that is, there will be a simultaneous flexion and adduction. The adduction of the thumb has no supplemental movements. Therefore, its paralysis, with inability to adduct, is to be considered as the sole pathognomonic sign of an ulnar nerve paralysis (Tinel).

Sensibility disturbances, hypesthesia, in case the nerve is injured higher up (on the forearm, for instance), will be confined to the volar and dorsal aspects of the hand. In the former, it almost reaches the middle line, involving also the little finger and the ulnar portion of the ring finger. On the dorsal surface, it affects the dorsal aspect of the hand to the middle and the dorsal surface of the last two and one-half fingers. As in median and musculospiral paralysis, the area of anesthesia does not coincide with the anatomic distribution of the sensory branches, being overlapped by those of the neighboring nerves.

Trophic disturbances are dryness of skin, desquamation, ulcers, blisters, etc. An incomplete lesion of the ulnar nerve causes so-called irritation symptoms—severe pain along the nerve, spontaneous or upon pressure; hyperesthesia and muscular sensitiveness and fibrous contractures of the hands, producing main en griffe. The tendons of the muscles become fibrous, and adherent to the synovial membranes, and the palmar aponeurosis thickens. The causalgia syndrome may occur as in median nerve lesions.

Prognosis, course and **treatment** are the same as those mentioned in the discussion of other nerve conditions.

Lumbar Plexus.—The lumbar plexus is formed by the first, second, third, and a portion of the fourth lumbar nerves. This plexus, placed within the psoas muscle, gives off several large nerves: Ilio-inguinal, iliohypogastric, genitocrural, obturator, external cutaneous femoris, and anterior crural. Isolated lesions of the lumbar plexus rarely occur, but have been described in simultaneous lesions of the sacral plexus as lumbosacral plexus paralysis. Of the nerves emanating from the lumbar plexus, only the obturator, external cutaneous femoris, and anterior crural have some clinical significance, though their isolated paralyses are very rare, especially in children.

The **obturator nerve** is formed by the anterior divisions of the second, third, and fourth lumbar nerves, passes between the psoas muscle and the body of the fifth lumbar vertebra, and supplies the adductors of the thigh (adductor longus and brevis muscles, musculus gracilis, adductor magnus, obturator externus). Lesions of the vertebral column, pelvic tumors, caries of the lumbar vertebræ, obstetric injuries in adults may cause paralysis of this nerve. Finally, it may be involved in the clinical picture of lumbosacral paralysis.

Symptoms.—The adductors of the thigh are involved in lesions of this nerve. The thigh cannot be adducted; the patient is unable to cross his legs, and the external rotation of the thigh is impaired.

Nervus cutaneus femoris externus is an exclusively sensory nerve, principally supplying the upper and external aspects of the thigh. Springing from the second or third lumbar nerves, it runs along the outer border of the psoas muscle, and, after leaving the pelvis, beneath Poupart's ligament, it enters a fibrous canal formed by a duplication of the fascia lata. Pelvic lesions, as in the foregoing nerve syphilis, vertebral lesions, infectious diseases (typhoid, influenza, measles), direct injuries to the nerve in its extrapelvic course, intoxications (lead, alcohol), sedentary habits, and diabetes may cause an inflammation of this nerve, a neuritis with a characteristic clinical syndrome known as meralgia paresthetica (Bernhardt-Roth's paresthesias[48]). The patient complains of paresthesias in the upper-outer, or antero-external, surface of the thigh. The paresthesias—numbness, tingling, sensations of freezing, burning, stabbing—increase with active movements, such as prolonged walking, marching, or standing, and subside with rest, or when the recumbent posture is assumed. In some cases there is actual pain (Sabraséz and Cabannes[49]) in the affected region.

Objective examination reveals diminished or lost sensation in the area covered by the paresthesia. Either all the qualities of sensibility are lost or diminished, or only those for pain, temperature, and touch.

Anatomic changes were found in the form of neuritis, perineuritis, varicosities, and nerve thickenings.

In patients under 28 years of age, paresthetic meralgia is quite rare, and so far has not been described in very young children. Musser and Sailer[50] found but one case in patients between the ages of 10 and 20 (out of 99 cases), while one of Roth's patients was 14 years old at the time of observation and Stembo's[51] was 16 years old. As the disease usually begins many years before the symptoms are pronounced, it must be admitted that it does occur in quite young patients.

Prognosis is good, though the disease may for last years and subside without any treatment. In obstinate cases, resection of the nerve may be indicated. Otherwise, rest, massage, and electricity suffice.

The nervus cruralis is formed by the posterior divisions of the second, third, and fourth lumbar nerves, their anterior divisions giving rise to the obturator nerve. From the groove between the psoas and iliacus muscles, the nerve passes to the thigh beneath Poupart's ligament, supplying the iliopsoas, sartorius, vastus internus, vastus externus, and rectus femoris muscles (i.e., the extensors of the leg), and, with sensory branches, the anterior part of the thigh as far as the knee and the anterior and inner parts of the leg (through the long saphenous nerve).

Causes.—Pelvic or vertebral disorders, lesions of the femur, and *especially of the knee joint*, are frequent causes of crural nerve paralysis. As the nerve has practically the same source as the obturator, they are usually affected simultaneously.

Symptoms.—The cardinal symptom of crural paralysis is paralysis of the leg for extension, also the inability to flex the thigh over the abdomen (ileopsoas). The paralysis is followed by marked and visible atrophy of the anterior thigh muscles, loss of knee-jerk, disturbances of walking on account of deficient extension of the affected leg. Knee-joint lesions, even the mildest injuries, may be the cause of the atrophy of the thigh muscles, but the manner in which the atrophy sets in is by no means settled. Some authors, as Vulpian and

Charcot, consider the atrophy and the nerve involvement as the result of reflex action on the anterior horn cells of the spinal cord, others consider it as the result of an ascending neuritis.

As in the obturator nerve, it may be a sign of lumbosacral plexus paralysis.

Sensory disturbances (anesthesia and hypesthesia), in complete lesion of the crural nerve, are confined to the antero-internal surface of the femur and inner surface of the leg and foot (down to the great toe).

The *diagnosis* of crural nerve paralysis should be differentiated from anterior poliomyelitis, in which sensory symptoms, as a rule, are absent, and from the so-called arthritic muscular atrophy, resulting from knee-joint trouble. In the latter, sensory disturbances are also absent.

Prognosis and *treatment* depend upon the etiology. The arthritic atrophies and paralysis do not yield entirely to treatment, but may improve somewhat following the use of electricity and massage.

Sacral Plexus.—The sacral plexus formed by the lumbosacral cord, which comprises a portion of the fourth lumbar and the entire fifth lumbar nerve, and the upper three sacral nerves, lies in the pelvis over the pyriform muscle. The plexus forms, at the margin of the great sacrosciatic foramen, into a single nerve, known as the great sciatic nerve, which with the pudic nerve is the terminal branch of the sacral plexus. Smaller or collateral branches are given off for the gluteal muscles, quadratus lumborum, pyriformis, and the internal obturator. The main and the most important branch is the great sciatic nerve. It commences as such at the lower border of the great sacrosciatic foramen and runs along the thigh, in the middle of which it terminates by dividing into the external and internal popliteal nerves. The external popliteal gives up a large nerve, the anterior tibial or peroneal, which supplies the extensor muscles of the foot and toes, while the internal popliteal gives off the posterior tibial for the flexors of the foot and toes. In short, through the external and internal popliteal nerves and their branches, the sacral plexus supplies almost the entire musculature of the lower limbs, except those muscles which derive their innervation from the lumbar plexus.

Paralysis of the sacral plexus, contrary to that of the brachial plexus, is very uncommon, being analogous in its symptomatology to that of a total paralysis of the great sciatic nerve. In children, paralysis of this plexus, aside from the possible pelvic or vertebral lesions, is often caused by tearing of the nerves or their roots during a so-called bloodless reduction of a congenitally dislocated hip joint. In these injuries to the sacral plexus, the lumbar plexus participates, the resulting clinical picture being that of lumbosacral paralysis.

Symptoms.—The entire musculature of the lower limb may be paralyzed, followed by atrophy, electrical changes, flaccidity, and loss of reflexes. Thus there will be foot-drop, inability to extend, flex, abduct or adduct the foot, inability to flex, extend, abduct or adduct the thigh, to cross the leg or rotate it. Cases of complete or incomplete lumbosacral paralysis have been reported by Lorenz,[52] Schlesinger,[53] Taylor,[54] Bernhardt,[55] Schuster,[56] Bade,[57] Peltesohn[58] and others. The most recent case was published by Levy, Tupper, and myself.[59] The patient, 19 years old, had fallen 15 years previously, with the legs widely abducted. After being confined to bed for nine months with a complete paralysis of the left leg, he began to walk. He always noticed weakness and smallness of the affected limb, which, however, did not cause him much discomfort. Eleven years after the injury he started to limp badly; the weakness became worse; he could walk only with great difficulty, bending his body forward and to the left, and throwing the leg forward. The entire left leg was markedly atrophied. The foot drooped and could not be extended or flexed. The leg could not be adducted or extended. The condition of the affected limb, of the opposite thigh, and the posture of the body are well shown in Fig. 51. In our case, sensibility disorders were very mild (in the foot and lower portion of the leg), appearing as patches of hypesthesia and anesthesia, but in the cases of Bernhardt and Schlesinger they were quite marked (outer and posterior calf muscles, external surface of feet and legs).

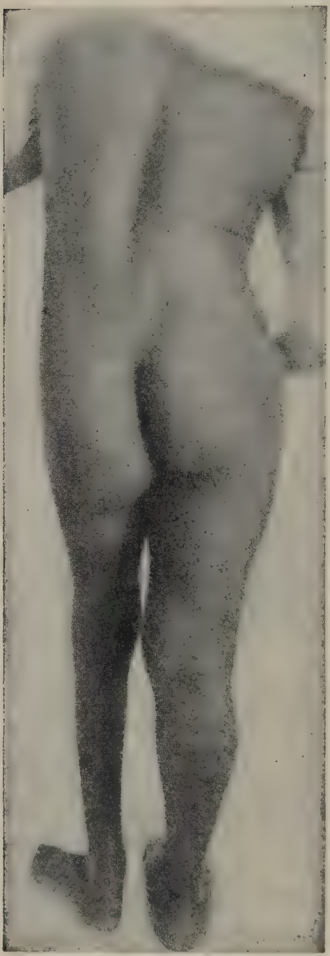

Fig. 51.—Lumbosacral paralysis, atrophy of the entire left lower extremity; tilting of the pelvis; bent trunk; scoliosis.

A sacral plexus lesion much more commonly results in a partial paralysis, which usually affects the *peroneal* nerve. In other words, paralysis of the peroneal nerve may be the sole manifestation of a sacral plexus or sciatic nerve paralysis. Even in the previously mentioned paralyses from operative orthopedic procedures, the lesion frequently resulted in a peroneal paralysis only. Calot explains this strange phenomenon as being the result of the pressure of the nerve against the fibula; Hoffman, as being the result of its lying in a poorer blood supply; Lorenz, by the fact that the nerve is immobilized and fixed against the head of the fibula. At any rate, the peroneal nerve is usually specially involved when the sacral plexus or the sciatic nerve is affected. Toxic, infectious, and other causes (traumatic—as fracture) may also involve the peroneal nerve.

Symptoms of Peroneal Nerve Paralysis.—As this nerve supplies the extensors of the foot and toes (tibialis anticus, extensor digitorum communis longus and brevis, extensor hallucis longus, peroneus tertius), its lesion will cause their paralysis. The result is a classical syndrome; dropfoot, paralysis of the basal phalanges, inversion and adduction of the foot, and steppage gait. In walking, the patient strikes the floor with his toes. In order to avoid this he raises the paralyzed foot higher than usual, putting it down toes first. In long-standing paralysis of this nerve, the condition of varo-equinus obtains. The peroneal paralysis is atrophic and flaccid, with electrical degenerative changes, and is usually associated with anesthesias. These, in total paralysis, are present over the anterior surface of the tibia and the dorsum of the foot and toes.

The condition must be differentiated from poliomyelitis, which is not difficult, if the anesthesias and a definite etiology are present. Otherwise it may be impossible.

The course, prognosis and treatment depend on the etiology.

The posterior tibial nerve supplies the flexor muscles of the foot and toes, the tibialis posticus, flexor digitorum communis longus, flexor hallucis longus, and all the small muscles of the foot through its terminal branches—plantaris, internus, and externus.

In its modes of innervation, the internal plantar is analogous to the median on the hand; the external plantar is similar to the ulnar. Thus, the former supplies the muscles of the great toe (except its adductor), abductor hallucis, flexor hallucis brevis, flexor brevis digitorum, the first lumbricalis, and the four digital branches of the inner four toes.

The external plantar supplies the abductor minimi digiti, flexor minimi digiti, three outer lumbricales, three interossei, adductor hallucis, and the skin of the toes (all or only four). Paralysis of this nerve may be caused by traumatic, toxic, or infectious causes, and results in paralysis of the muscles supplied by this nerve, i.e., of the flexors of the foot and toes. Both appear dorsally extended and abducted from the action of the peroneal nerve. The foot is inverted, the external border everted or raised, and the toes are claw-like (from the paralysis of the interossei and lumbricales). Long-standing paralyses cause a club foot deformity, pes calcaneus. As in the upper extremity, supplementary movements may take place in the paralysis of the muscles of the lower extremity.

Sensibility disturbances are confined to the postero-lateral surface of the leg, outer edge of foot, sole, and plantar surface of the toes.

A *combined paralysis* of the peroneal and posterior tibial nerves occurs in complete paralysis of the sciatic nerve, sacral plexus, or of the roots (motor and sensory) which give rise to the nerves forming the lumbosacral plexus, that is, the cauda equina. The etiology, symptoms, and course of complete sciatic paralysis are the same as in sacral plexus paralysis, while lesions of the cauda equina are usually considered in the discussion of spinal cord diseases, and cannot, therefore, be discussed here in detail. Only points of differential diagnosis from sacral plexus or sciatic lesions will be described here briefly. The cardinal features of cauda equina lesions are pain and muscular atrophy, with paralysis confined to both lower limbs, mostly asymmetrically. The pain is very severe and continuous, with exacerbations. It involves the entire extremity and may be more severe on one side than on the other. The muscular wasting and paralysis may affect the entire limb, or only some of the muscles, depending upon the seat and height of the lesion. Since the paralysis is asymmetrical, one limb shows more muscles involved than the opposite one. The pain and atrophy are associated with anesthesias, or hypesthesias, which are of a radicular type, for they follow the distribution and course of the roots, and not of the peripheral nerves as in sciatic nerve lesions. Electrical changes are less pronounced than in the peripheral type, being usually partial. The severe pain, irregularly scattered atrophies and sensory disturbances, combined with other features which cannot be discussed here, are usually sufficient to permit a diagnosis of a cauda equina lesion.

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CHAPTER CLXXIV

HEAD SHAKING WITH NYSTAGMUS IN INFANTS

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Nomenclature.—Various names have been given to this condition. Those which include the word spasm, such as spasmus nutans, nutatio capitis spastica, gyrospasm, nodding spasm, are objectionable, because it is not a spasm in the sense in which that term is usually used. The terms gyrospasm, rotatory spasm, head nodding and nodding spasm are inaccurate, because the movements may be in any direction. The term spasmus nutans has sometimes led to confusion with the condition known as eclampsia or epilepsia nutans, which is an entirely different disease. Salaam tic, a term sometimes used by French authors, is objectionable because this condition has none of the characteristic features of a tic. Everything considered, head shaking with nystagmus, seems to be the least objectionable name.

Historical.—This condition was first described by Ebert[1] in 1850, and in 1851 by Romberg and Henoch.[2] In 1890 Hadden[3] published two important papers, with a report of 21 cases. Caillé[4] of New York was the first to note that when the nystagmatic eye or eyes were bandaged, the movements of the head ceased. In 1897 Raudnitz[5] published his exhaustive monograph on the subject, based on 15 cases, in which he particularly emphasized the importance of the dark room in the etiology of this condition. In 1911 he published a report on the producton of this condition experimentally in young dogs. In England and Scotland, Still[6] and Thomson[7] have reported large series of cases. In this country Peterson,[8] Aldrich,[9] Abt,[10] Judson,[11] Miller,[12] Hamill and Posey,[13] Hill[14] and others have published cases.

Frequency.—This condition seems to be more common in some countries than in others, if we may judge by the number of reported cases. It is certainly more frequent in large cities. In Budapest at the children's clinic, 15 cases were observed in three years among 52,215 patients; in Prague, eight cases in two years among 1223 patients; in Edinburgh, 35 cases in 11 years among 21,752 patients; in Basel, two cases among 6154 patients; in Christiania 10 cases were seen at the children's clinic in 10 years. The disease appears to be not uncommon in London, Edinburgh, Berlin; and in this country in New York and Philadelphia. During the last 19 years I have had 64 cases under my observation at various children's clinics in New York. The first 15 were seen during 1899, 1900, 1901, at the Good Samaritan

Dispensary, chiefly in the service of Doctor Koplik. These figures would indicate that this condition is met with in New York City in about one in every 700 infants coming for treatment. The number of cases varies markedly in different years. In 1901 I saw eight cases; in 1906, nine cases; in 1910, 13 cases, and in 1918, seven cases; whereas in some years, 1904, 1908, 1909, 1912, 1914, 1916, no cases were seen. This difference in the prevalence of the disease in different years has also occurred in other cities; for example: In Budapest, in 1891, there was one case in 17,400 patients; in 1894, nine cases in 16,563 patients. It is relatively somewhat more common among negro children than among white children in New York. In 1910 I saw 10 cases in negro children at the Vanderbilt Clinic. This is probably due to the poor hygienic conditions and the greater prevalence of marked rickets in these children.

Sex.—Females are slightly more frequently affected. In my series, there were 29 males, 35 females. In 133 previously reported cases there were 62 males and 71 females.

Seasonal Incidence.—This condition shows a very distinct seasonal incidence, as shown in the accompanying tabulation:

OCCURRENCE OF HEAD SHAKING WITH NYSTAGMUS BY MONTHS

	Jan.	Feb.	Mar.	Apr.	May	June	July	Aug.	Sept.	Oct.	Nov.	Dec.
Previously reported 110 cases.......	37	16	10	1	1	1	1	2	4	6	8	23
Herrman, 60 cases...............	20	12	6	1	2	0	1	2	2	3	4	8
Hours sunlight, Edinburgh (Thomson)........................	34	57	98	123	165	157	138	145	104	83	47	24
Hours sunlight, average, 1911–1917 New York (Herman)...........	140	177	223	220	270	288	279	265	243	200	177	151
Mean temperature, average, 1911–1917 New York (Herrman)......	33.5	29.7	37.2	49.7	60.7	68.3	74.7	72.9	66.4	57.1	44.7	34.4

It will be seen that it is much more prevalent during the winter months, nearly 70 per cent. of all cases occurring during December, January, and February. The largest number of these begin in January, and recurrences are almost always in this month. As Thomson has pointed out, the number of hours of sunlight, in temperate climates, is at its minimum during the winter months (Fig. 52), and this may bear a definite relation to the prevalence of the condition. It may also account for the very few cases reported in France, Italy, and Spain, for it seems very unlikely that the disease could be overlooked. However, it does not appear probable that the relation is a direct one, and there is no reason to suppose that the amount of sunlight bears a direct relation to this disease. It would be interesting to determine whether the head shaking is rare in countries in which there is an unusually large percentage of sunshiny days, for example: New Zealand, and whether the condition is prevalent in the months of December, January, and February in places in which the summer and winter seasons are different than our own. It will be noted (Fig. 53), that the incidence curve of head shaking also corresponds quite closely to the mean temperature curve, so that the temperature might also be important, insofar as during very cold weather the windows

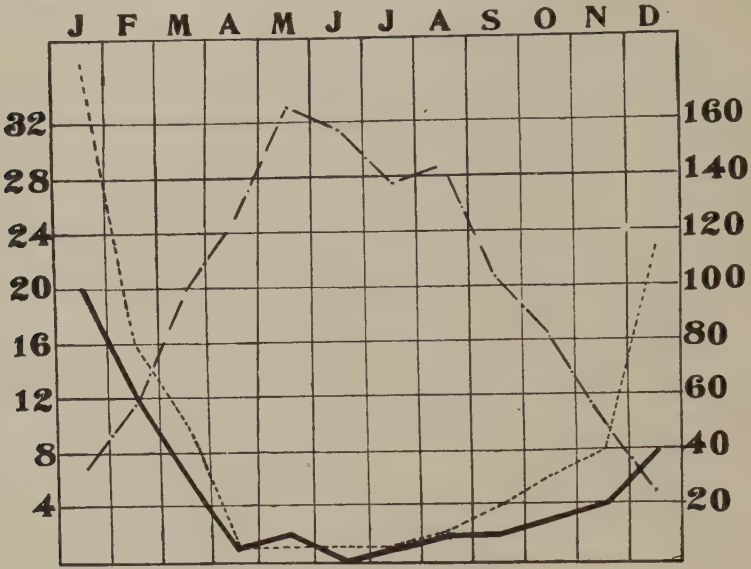

Fig. 52.—Seasonal incidence of head shaking with nystagmus, ——— sixty-four cases observed by the author, ... 110 previously reported; —.—.—. number of hours of sunlight each month in Edinburgh. (Thomson.)

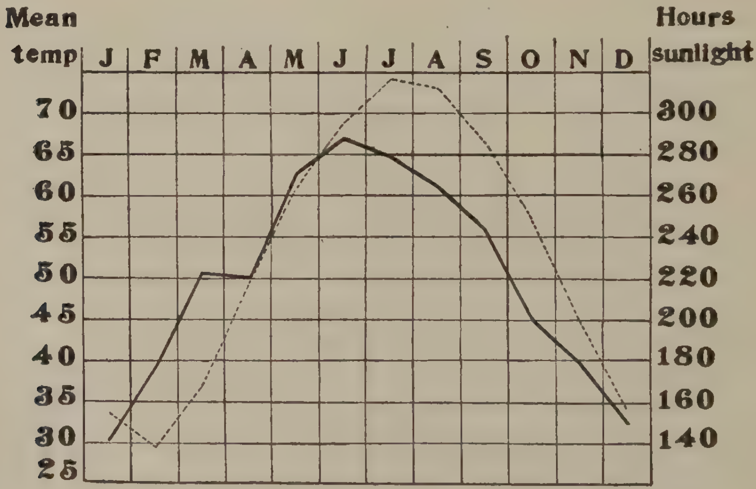

Fig. 53.—New York City, 1911–1917; ——— number of hours of sunlight; . . . . mean temperature.

would be closed and the infants not be taken out as much as usual. This may possibly account for the unusually large number of cases seen during the past winter (1917), which was an extremely severe one.

Age Incidence.—The disease is most common between 4 and 12 months of age, 75 per cent. of all cases occurring at that time. In my series the age of onset could be definitely determined in 61. It was one at one month, two at two months, two at three months, two at four months, three at five months, eight at six months, eight at seven months, five at eight months, five at nine months, six at 10 months, four at 11 months, six at 12 months, and one at four years. The youngest in the series was one month old. Schoenberg[15] has also reported a case at that age, and Gee[16] one at six weeks of age. After the eighteenth month a first attack is rare, but there may be recurrences at that time. In some of the late cases, the original attack may have been overlooked. My oldest patient was four years of age. Henoch reported a case of three years, Still one of four years, and Mowat[17] one of five years. It is pretty well established that head shaking is a developmental disease, and that it occurs in certain predisposed infants at a time when the associated movements of the head and eyes are being perfected.

Etiology.—Family History.—In very few of my series was there a distinct neuropathic family history. In two instances the parents were cousins. In four cases the mother was extremely nervous, in three markedly anemic; one had frequent attacks of migraine; one had hysteria. In five families one or more of the other children had convulsions; in one family one child had epilepsy, and in another family a child died of meningitis. In one instance the condition occurred in twins, and in another family two children were affected. Henoch and Reyher also observed the disease in twins; and Thomson and Hadden have reported instances of more than one case in a family. Three of my cases occurred in mongolian imbeciles. Unlike the latter disease, head shaking does not attack more frequently the first or last child, and it is not more common in families in which there are a large number of children.

Previous History.—In three of my series, the labor was difficult and the delivery instrumental; one was a "twilight" baby, and one was prematurely born. None of these factors seems to play a part in the causation of head shaking.

Feeding.—At the onset of the disease, 35 of my patients were breast fed, 18 artificially fed, and 11 were on mixed feeding, so that the character of feeding has no direct relation to the disease. As one would expect, the rachitic manifestations when present were more marked in the artificially fed infants, so that to a certain extent such infants were somewhat more predisposed to head shaking.

Previous Disease.—Illness, by lowering the vitality, apparently is an etiologic factor in those infants who are already predisposed. Five of my cases followed an attack of bronchitis, three followed pneumonia, six patients had digestive disturbances, two measles, and one each naso-pharyngitis and otitis. Intercurrent disease tends to increase the head and eye movements.

Teething.—It is to be expected that this condition, which plays such an important etiologic rôle in the minds of mothers, should also be held responsible for head shaking. It would not be surprising if the nystagmus were ascribed to the eruption of the "eye" teeth. Ebert, Henoch, and Still believe that the eruption of teeth often plays an important part as a source of peripheral irritation. In exceptional cases this may be so, but I have not been able to convince myself that the rôle is an important one. In 18 of my cases the onset of the disease antedated the eruption of teeth. Ebert, who first described the condition, writing in 1850, says: "It is well known that febrile and spastic conditions which are related to the eruption of teeth may antedate the actual appearance of the teeth." If we accept this statement, then anything is possible. I recently saw an infant in whom the eruption of two incisors occurred on the day on which a measles eruption appeared. To the lay mind it seems like cause and effect. In my patients teeth appeared during the course of the disease, but I never noticed an exacerbation of the symptoms at that time. Fatigue does tend to increase the symptoms, so that it is possible that a fretful child that is teething may sleep less, and in that way the head movements might become more distinct. Other sources of peripheral irritation, worms, otitis, etc., have been mentioned as causes, but the evidence is not conclusive.

Trauma.—*Fall.*—In a small percentage of cases this may reasonably be considered an exciting cause in a predisposed child; but it must be remembered that a large number of infants fall and very few show any permanent injury. Only when the head shaking begins shortly after the fall can a relation be claimed; and it is also possible that the fall is a symptom of an already existing co-ordination neurosis rather than its cause. In six of my series there was a definite history of a fall just preceding the onset of the disease. In one of Hadden's patients two relapses occurred following falls on the head. Peterson[8] says: "Perhaps a concussion of some kind may be found in most of the cases to be the determining factor—a concussion setting up in the centers of the spinal accessory and at times in those of the oculomotor nerves an arrhythmic discharge."

Hygienic Conditions, Social Position, Location of Rooms.—The hygienic conditions and social position of the family play an important part. Many authors say that the disease does not occur among the well-to-do. This is not absolutely correct, as I have had two cases in families in comfortable circumstances, and Still mentions having seen three such cases. It is rare, however, among this class compared with the frequency with which rickets, laryngismus stridulus, tetany and convulsions occur in them. In large cities, head shaking is more common among those who live in the poorer tenement districts. In my series 11 families lived in the basement, 20 on the first floor, nine on the second floor, 12 on the third floor, seven on the fourth floor, and five on the fifth or top floor. It will be noted that one-half of the families lived in the basement or on the first floor; those living in the basement were not only the poorest, but their rooms were dark and ill ventilated, and the intelligence of the mothers was lower than the average.

Dark Rooms.—In 55 of the cases of my series the character of the rooms could be accurately ascertained. I visited nearly all of them myself. It is important to investigate oneself and to determine the exact conditions during the months preceding the onset of the disease. The crib may be placed in a dark corner of a room not otherwise very dark; the shades may be drawn down a large part of the day so as to darken the room and favor sleep, or the crib may be curtained in order to prevent the light from shining in the infant's eyes. In 40 of my series, the infant was more or less in the dark, and in a large number of these cases it was necessary to use artificial light in the afternoon during the winter months. In the remaining 15 the room and the spot where the infant lay were fairly light, and in a few of these the room was bright even during the winter months. In several cases, although there was a light room, the infant was kept most of the time in the kitchen, a dark room, so that the mother could watch it while cooking; in several others it was impossible for the mother to take the baby out on account of sickness or the large number of other small children that required attention. On the other hand, in a few cases the mother definitely said that the baby was taken out regularly. It must be remembered that in certain crowded sections of the city a large number of people live in dark rooms, so that only a small portion, probably not more than one in 100 infants living in such rooms, contracts the disease. Here again we must assume an individual predisposition. The same has been noted in so-called miner's nystagmus; only 3.5 to 4 per cent. of all coal miners working under exactly the same conditions are affected. There can be little doubt that the dark room is an important factor in a large number of the cases of head shaking. During the last 11 years I have seen the same class of patients at the Lebanon Hospital out-patient department in the Borough of the Bronx as I saw during 1899–1902 at the Good Samaritan Dispensary on the Lower East Side of New York. Making due allowance for the difference in the number of patient, the disease was more common in the latter section, principally, I believe, because in the Borough of the Bronx the majority live in rooms which are better lighted and better ventilated. A series of four cases of head shaking reported by Rietschel[18] from Finkelstein's service is interesting. On account of the suspicion of communicable disease these four infants were transferred from the well-lighted ward of the infant asylum to the dark room of a barracks. Shortly after, they developed head shaking. One of the infants was returned to the well-lighted and well-ventilated ward of the asylum and the head movements disappeared. Raudnitz was able to produce experimentally a similar head shaking with nystagmus in puppies by placing them in a dark cage at or very shortly after birth.

In from six to eight weeks they developed nystagmus, and after three or four months, shaking of the head. If they were placed in a darkened cage after they were two months old, these symptoms did not develop. Here again we note that the causative factors act only at a certain stage in the development of the nervous mechanism which controls the associated movements of the head and eyes. As in infants, at first the nystagmus was only seen when the animal looked in certain directions or when the head was held, and the head movements ceased when the eyes were bandaged. If the animals were placed in the open a few days after the nystagmus first showed itself, it disappeared in from one to two weeks, and if they were returned to the dark cage, the nystagmus reappeared in from two to seven days. If the animal was kept in the dark, however, so that the nystagmus persisted for several months, it required as much as nine months' exposure to daylight to cause the nystagmus to disappear. The examination of the fundus, the vision, and the microscopic examination of the brain and retina of the affected animals showed no pathologic changes. It is worth noting that in Raudnitz's experimental work on animals the cage was dark and apparently there was no artificial light to which the eyes of these young animals was attracted. In explaining the causation of the condition in infants he laid stress on the straining of the eyes in looking at an artificial light some distance above the level of the head. Raudnitz believes that the nystagmus associated with the head shaking in infants is analogous to that observed in coal miners. Although there are some points of analogy, there are some objections to this theory. The nystagmus of coal miners seems to be due to the strained position of the head, eyes, and body of the men, working in a place which is imperfectly lighted. The darkness alone is not sufficient, for this form of nystagmus occurs only in coal miners. The strained position of the head and eyes alone is not sufficient, for it does not occur in ceiling painters, and in others who have to look more or less constantly upward. A large percentage of the miners affected have errors of vision, while the infants with head shaking associated with nystagmus have no such error. The nystagmus of miners is usually bilateral and conjugate, that of the infants is usually unilateral, or is much more marked in one eye, and is frequently not conjugate. In miners head movements are uncommon and are only present in severe cases; in infants head movements are the rule. In miners the head movements do not cease when the eyes are bandaged as they do in the infantile form.

Rickets.—Kassowitz,[20] Hochsinger,[21] and Stamm[22] consider the head shaking of infants a rachitic manifestation. In my series, 45, or 70 per cent., of the patients showed some manifestations of this disease. This is about twice as frequent as in other infants brought for treatment. Thomson's figures are 94.2 per cent. rickets in the affected infants, 46.9 in other infants. Of course, the percentage of rickets will vary according to the clientele. For example: In New York City rickets is much more common in negro children, so that it is not surprising that every one of my 10 cases of head shaking in negro children showed distinct rachitic changes.[23] However, in 19 of my cases rickets was not present, so that it cannot be an indispensable factor. Several other points indicated by Thomson are of interest. In head shaking, anti-rachitic treatment is not followed by the rapid improvement which we are accustomed to see in rickets. This has been my own experience and is contrary to that of Stamm, who noted distinct improvement in the head shaking in a few weeks. Cases of head shaking are rarely associated with laryngismus stridulus, facial irritability, tetany, or convulsions. In my series, although many had marked rachitic manifestations, only three had laryngismus stridulus and facial irritability, and in Thomson's 35 cases only two had laryngismus stridulus and facial irritability. There does not seem to be a distinct relation between the frequency of laryngismus stridulus and head shaking in a given locality. In Vienna, Kassowitz in six years saw 370 cases of laryngismus stridulus, that is, on the average 61 cases a year; in Edinburgh, Thomson in three years saw only 61 cases, that is, 20 a year. On the other hand, Kassowitz in eight years saw only nine cases of head shaking; that is, about one a year, while Thomson saw 35 cases in 11 years; that is, three a year. If these conditions were due, as Kassowitz supposes, to the increased vascularity of the bones of the skull and the neighboring tissues, then we should expect that head shaking would be frequently present in rachitic infants with craniotabes; however, such is not the case. Rietschel tested the electrical reactions in four cases of head shaking, and three of the four gave normal reactions. While head shaking is rare in well-to-do families, rickets, laryngismus stridulus, tetany, and convulsions are not uncommon. As is well known, rickets involves not only the bones, muscles, and ligaments, but the central nervous system as well, so that it is not surprising that a rachitic infant should be somewhat more predisposed to the development of a functional neurosis.

Pathogenesis.—Notwithstanding the great amount of investigation on this subject, the exact explanation of the disturbance of the nervous mechanism which results in the head and eye movements is still something of a puzzle. The solution requires the combined investigation of the pediatrist, neurologist, and ophthalmologist. Thomson says: "The nervous mechanism whose function is at fault is evidently that which includes the semicircular canals in its circuit, and which regulates the co-ordination of the movements of the eyes with those of the head. Dr. Alex Bruce tells me that the nucleus of Deiters (one of the end nuclei of the vestibular nerve, lying at the lateral angle of the fourth ventricle) is in all probability the seat of the disturbance. He infers this from its connection with the oculomotor nuclei and with the anterior cornua of the cord."

Henoch had about the same in mind when he wrote: "The frequent combination of head shaking with nystagmus is interesting, as it indicates that the nuclei of the spinal accessory and the upper spinal nerves which supply the affected muscles of the head and neck, are in close relation to those of the eye muscles."

The most satisfactory explanation of the pathogenesis of this condition is that originally suggested by Hadden and later modified and amplified by Mills[24] and Aldrich. Mills says: "Hadden believes that the symptoms point to an instability of the motor centers above the nuclei in the spinal cord and fourth ventricle, and he would therefore attribute the disorder to a functional or other disturbance of the cerebral cortex. The child has acquired certain voluntary or purposive movements of the head and eye-balls, but these have not as yet become thoroughly organized and fixed in the psychomotor areas of the brain, hence, a dissolution takes place because of the instability of the strained cortical centers to stand the work to which they have been too early subjected."

Aldrich amplifies this as follows: "The writer in considering the pathology of these cases accepts as a fact the obvious relation of the head and eye movements, hence we have to consider an automatic head and eye movement in a child of unstable nerve development, and at an age in which the associations of those necessary movements are being educated and fixed in the psychomotor areas of the brain. Anyone that has observed the head and eye movements of the infant cannot have failed to mark the inco-ordinated attempts at associated movements of the head and eyes. The fibrous connection between the visual areas of the cerebral cortex and the fovea centralis that is necessary to perfect vision does not fully develop until after birth, and only gradually becomes medullated and fit for function. After this process is completed, which takes place only when the cortical cell has attained a degree of maturity that provides ample nutrition for its axis cylinder, then the structural and physiologic integrity of the central neuron is complete. During this time the same process of development in the peripheral neuron is going on with even greater rapidity, and we have the beginning of one of the most wonderful and mysterious processes of the nervous system—the communication between the central and peripheral neuron . . . Binocular adjustment, judgment of fixed objects, spatial relations, including station, are acquired by the child as the medullation progresses. If this progress of medullary development of the nerve supply of the eyes is either interfered with by disturbance of nerve nutrition or rendered functionally imperfect through early acquired opacity of the refracting media, or by congenital absence of a place in the retina that has more acute and perfect vision than elsewhere (fovea centralis), or a congenital defect in the receptive center in the cortex, or an exhaustion of any of that delicate apparatus, then we are liable to get a rapid to and fro movement of the eyes—nystagmus."

Symptoms.—Movements of the Head.—The movement of the head is usually the first manifestation noticed by the mother, but in a certain number it may be preceded by a peculiar "cocking" of the head to one side. In some the nystagmus may precede the head movements but may not be noticed because it is present only when the child looks in certain directions, or when the head is held. The muscles chiefly affected are the sternocleidomastoid, the trapezius, and the rectus capitis. The movements are regular and rhythmical and from 60 to 100 a minute. Usually they are not continuous, but come on at longer or shorter intervals. The direction is rarely purely horizontal, vertical, or rotary, but when primarily horizontal, slightly upward to one side and downward to the other, the excursions often being greater to one side. If primarily vertical, it is rarely a pure nod forward and backward. The movement may be a combination of the horizontal and vertical, but they may be in different directions at different times. In my series the movements were primarily horizontal in 44, vertical in 12, rotary in four, and horizontal or vertical at different times in four. The movements are often more distinct when the child is angry or fatigued, and during an attack of some intercurrent disease. They cease when the child is lying down and in sleep. There is, however, an occasional exception to this rule; for example: In one of my cases, in which the movements were unusually frequent and well marked, they continued when the child was lying down, and Amberg[25] also mentions a case in which the movements continued during sleep. It was first noticed by Caille, and later by Raudnitz, that the movements of the head ceased when the eyes were bandaged. It is not possible to test this in all patients, because some object so strongly that the movements of the head due to anger prevent the drawing of definite conclusions; but in 14 of my cases, I was able to corroborate this statement. The movements of the head are best seen when the infant is gazing in the distance and is not interested in any particular object. If an object is brought within a short distance, so that it is definitely fixed, the movements of the head cease.

Movements of Other Parts of the Body.—I have not observed movements of the extremities, such as described by Thomson, Still, and others, or of the trunk, as reported by Hadden. Ausch[26] noticed in one of his cases movements of the hands, as if the child was vexed. I have frequently seen such movements in infants who were entirely free from head shaking.

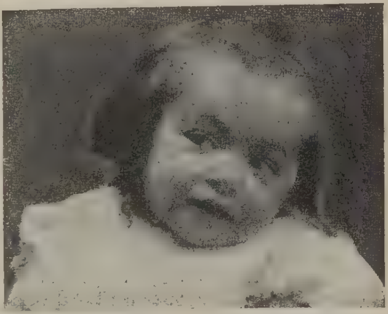

Fig. 54.—Peculiar cocking of the head seen in infants affected with head shaking with nystagmus.

The Peculiar Position of the Head (Fig. 54).—About one-half of these patients have a peculiar way of "cocking" the head to one side and looking out of the corners of the eyes. This may precede the head movements and may persist after these movements have ceased. In 75 per cent. of the infants in whom this "cocking" was present the head was inclined to the left. This is noteworthy in connection with the fact that in 16 of the 23 cases in which the nystagmus was unilateral, the left eye was affected. Some of the patients have a habit of looking down with coy expression; others throw the head back and look out of the partially closed lids.

Movements of the Eyes.—The nystagmus may appear before, simultaneously with, or after the head movements. It may persist after the head shaking has ceased. In 35 of my cases the nystagmus was bilateral, in 23 unilateral. Of the latter, 16 affected the left eye, seven the right eye. Even when the nystagmus was bilateral, it was frequently more marked in one eye. In six cases the nystagmus was not seen when the patients were examined in the ordinary way, but it is known that in some cases the nystagmus is so slight that it can only be detected by an ophthalmoscopic examination. The direction of the nystagmus is often not purely horizontal, vertical or rotary; it may be different in the two eyes, or when the patient looks in different directions. In the 58 cases in my series in which nystagmus was present, it was primarily horizontal in 36, vertical in six, and rotary in 15; in the remaining case it was impossible to determine the direction. The movements of the eye are much more rapid than those of the head, about 200 to 300 a minute. The nystagmus is increased by holding the head, and is more distinct when the child looks in certain directions, usually when it looks upward and to the right, when the right eye is affected, and upward and to the left, when the left eye is affected. It is also more distinct when the child fixed an object at a short distance, and when the eyes are converged. It is not present during sleep, as I was able to determine in several infants that I examined. When the nystagmus is bilateral it is not necessarily conjugate. According to Thomson,[7] it differs from other forms in that the pupils alternately approach and recede. Spicer[27] was not able to verify this in all of this cases. Thomson also says that in rotary nystagmus the center of the pupil does not describe a circle, but an ellipse or an irregular figure. The movements of the head are not simply compensatory to those of the eyes, for the rate is different, and the two are not necessarily in the same direction; for example: The movements of the head may be vertical, while those of the eyes are horizontal. The nystagmus may persist after the head movements have ceased, and it may recur without the appearance of head movements. In exceptional cases the nystagmus may be the only manifestation. Such cases may be regarded as atypical or incomplete. In four of my series, convergent strabismus was present; in three there was hippus; three had associated movements of the upper lids, "nictitation;" and in two there was some sensitiveness to light; 12 of my patients were examined by ophthalmologists, and none of these showed any changes in the fundus, defects of vision, or opacities in the media. Similar examinations reported by other authors were also negative, except that in two of Hadden's cases the fundus was light colored, and in one a slight crescent of atrophy was present around the left disc. In a few of my patients, I instilled homatropin in order to paralyze accommodation. The nystagmus was diminished, but the head movements continued. This is what one would expect, for as previously mentioned, the head movements cease when an object is distinctly seen and fixed; by paralyzing accommodation this becomes impossible. As nystagmus is sometimes associated with disturbances in the semicircular canals, Raudnitz had the ears examined in several cases. None showed any lesions, and syringing of the ears had no especial effect on the nystagmus.*

Associated Conditions.—In my series, six patients were anemic, five had an enlarged spleen, two tuberculous lesions, two papular urticaria, two eczema, three a geographical tongue, one blepharitis ciliaris. One child had attacks of apnea. Hadden reports cases with attacks of unconsciousness, partial or complete, and convulsions. Such cases are rare, and the more severe probably represent cases of so-called epilepsia nutans.

Duration.—The condition seldom lasts less than two, or more than 12 months. In my series, the duration of 50 cases could be determined. In six it was three months; in six it was four months; in 12 it was five months; in six it was six months; in four it was seven months; in four it was eight months; in two it was nine months; in four it was 10 months; in six it was 12 months. In three there was a recurrence during the following winter, two in January and one in February. In exceptional cases, however, the disease may last for more than one year. Spicer mentions a case in which it was present at 4½ years of age, and another in which the nystagmus persisted at 13 years of age.

Subsequent History.—I have been able to follow 15 of my cases for a period of from 2 to 10 years. Of these, eight had no subsequent disease that had any relation to the head shaking. Of the remaining seven, two had enuresis, one of these until she was six years of age; one had convulsions; one pavor nocturnus; one a facial tic; one was extremely sensitive and also anemic; and one was a very restless sleeper and had grinding of the teeth. With the exception of the three cases in mongolian imbeciles, all the children showed a normal average mental development. I recently saw a girl of 13 years with a mild type of exophthalmic goiter (Graves' disease) who had had head shaking in infancy. In none of the five children who were followed to the eighth year did chorea develop. This is of some interest, as several authors have thought that there was a possible relation between these two diseases.

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* Zappert, in a paper recently published, Med. Klin, xx (1924) p. 335, admits the importance of the effect of a dark room on the eyes, but points out that the condition occurs only in infants who have started to walk or were held upright, but never in those who were already walking, or still reclining. Thus the increased function of the vestibular apparatus may play an important rôle.

In this connection, I might mention that as early as 1900, in a personal communication, Professor Fronz, of Vienna, wrote to me, "head shaking with nystagmus belongs to the automatic movements caused by a feeling of dizziness, which is in turn due to irritation of the floor of the fourth ventricle. It is also possible that in some cases there is a disturbance in the labyrinth."

Diagnosis.—The principal features of this condition are so characteristic that the correct diagnosis is seldom difficult. Some of the earlier authors confused it with eclampsia or epilepsia nutans, which is really an epileptic equivalent. This disease occurs in attacks, and is associated with partial or complete unconsciousness. There is no nystagmus. In head shaking there are no sudden attacks, the movements are more or less continuous, and consciousness is not lost. The movements in head shaking are entirely different from those of a tic or habit spasm. They are smooth, regular, and rhythmical, not jerky. Tics do not occur at such an early age, and they are not associated with nystagmus. The rocking movements of the trunk and head, frequently seen in infants and young children, have only a remote resemblance to head shaking. The movements are voluntary, and even when they are not a manifestation of masturbation, seem to give the patients a certain amount of pleasure or satisfaction. Nystagmus is absent. Head shaking has occasionally been confused with the nocturnal movements of the head, Jactatio capitis nocturna, which is sometimes seen in older children. These movements differ from the head shaking of infants in that they occur during sleep, or at least in a subconscious state, and are not associated with nystagmus. Head rolling, like head shaking, is seen in rachitic infants, but it is only present when the child is lying down; it is a voluntary act, and it is not associated with nystagmus. There is a rare form of congenital head shaking with nystagmus which is due to pathologic changes in certain areas of the brain. This condition, as well as a number of other diseases of the central nervous system, associated with head movements or nystagmus, can easily be differentiated from the benign head shaking of infants, by the fact that the disease is progressive and persists throughout life. In all these conditions when nystagmus is present it is bilateral and conjugate.

Prognosis.—Head shaking with nystagmus in infants is a benign disease; all patients recover. The mothers may be assured that this disease will not interfere with the general health, or with the future mental and physical development of the child. The disease usually begins in the winter months and disappears during the following summer. Occasionally there is a recurrence of symptoms the following winter, but this also is of short duration. A few of the patients show a neuropathic tendency in later childhood.

Treatment.—The general condition, the hygienic surroundings, should be improved if possible. The room should be light, and the patient should be kept in the open air as much as possible. In several of my cases the mother noticed that there was a rapid improvement when the child was taken to the country. If there are any sources of peripheral irritation, these should be removed; if there are digestive disturbances, the diet should be regulated. Those infants that present rachitic manifestations should receive phosphorized cod-liver oil. In most cases this treatment is sufficient. I have not seen any marked improvement following the administration of bromids, belladonna, or arsenic.

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CHAPTER CLXXV

EXTERNAL HYDROCEPHALUS. ACUTE INTERNAL HYDROCEPHALUS. CHRONIC INTERNAL HYDROCEPHALUS

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Hydrocephalus, a descriptive term, is applied to those conditions in which there is an excessive accumulation of cerebrospinal fluid within the cranial cavity. It is either external or internal. Internal hydrocephalus may be acute or chronic.

External hydrocephalus is characterized by the accumulation of cerebrospinal fluid, external to the cerebral cortex. This form of hydrocephalus occurs in association with such developmental defects of the brain as an absence or an imperfect development of one or both hemispheres; it may be caused by the perforation of a cortex greatly thinned by internal hydrocephalus, in which case the cortex never collapses sufficiently to allow all of the fluid to become extra cerebral; and cases are described following meningeal hemorrhage, pachymeningitis and cerebral atrophy. Whether external hydrocephalus should be regarded as a clinical entity is open to question as its pathogenesis is not well understood. It is an exceedingly rare condition closely associated with and in all probability secondary to internal hydrocephalus. The majority of cases are found at the pathologic examination.

Acute Internal Hydrocephalus.—Internal hydrocephalus whether acute or chronic is always secondary to a primary condition interfering with the normal circulation and absorption of cerebrospinal fluid. Meningitis following an infection by the meningococcus or the tubercle bacillus is the most common cause of acute internal hydrocephalus; more rarely it follows meningitis due to the pneumococcus, the influenza bacillus, the streptococcus and the staphylococcus aureus. Tumors of the brain although comparatively rare in children may be the exciting factor. It is questionable whether other inflammatory conditions of the central nervous system, such as acute poliomyelitis, and encephalitis, ever produce more than a temporary and insignificant acute hydrocephalus. The symptoms and the course will be discussed in the chapters devoted to the complications of the diseases above mentioned.

Chronic Internal Hydrocephalus.—This is the most frequent form in infants and young children and therefore is of chief interest to the pediatrician. It is characterized by the progressive accumulation of cerebrospinal fluid in the ventricles, causing their dilatation, a compression and thinning of the cortex and enlargement of the head. Chronic internal hydrocephalus may be either congenital or acquired.

Etiology.—Almost every conceivable cause, direct and indirect, has been held responsible for the development of hydrocephalus. Alcohol, rickets, trauma, tuberculosis, syphilis, heredity, lack of resistance of the brain tissue, osteogenetic defects of the skull and many other even less likely possibilities have been suggested. Alcohol and rickets have no direct bearing on the production of internal hydrocephalus. Syphilis and tuberculosis cause hydrocephalus only when the lesions are so located as to interfere with the absorption of the cerebrospinal fluid. I have seen only one case in which the hydrocephalus was proven to be the result of syphilis. A familial tendency to hydrocephalus is not common although instances are on record in which the condition developed in several members of the same family. The production of hydrocephalus by trauma is difficult to explain. I have observed one patient whose father, a physician, insisted that the onset of the disease dated from a severe fall.

The chief causative factors of internal hydrocephalus are meningitis, congenital abnormalities of the brain, and tumors. A large percentage of the cases are secondary to meningitis. The meningococcus in so far as is known is the only organism that causes a meningitis which pursues a chronic course or in which spontaneous recovery takes place. Therefore, when chronic internal hydrocephalus follows a meningitis, it is in the majority of instances due to the meningococcus. The symptoms of the meningitis are often indefinite so that an infection of the meninges preceding the development of the hydrocephalus may be obtained only by most careful questioning. In congenital hydrocephalus the existence of an intra-uterine inflammatory process has often to be assumed, as the same pathologic processes are found as in the acquired form. In the studies of Dandy and Blackfan the primary cause for the hydrocephalus in 21 cases as determined from the history and pathologic findings was as follows: Sixteen cases resulted from meningitis, three cases were due to a congenital absence of the aqueduct of Sylvius, one case was caused by syphilis and one case by a tumor of the brain.

Various congenital anomalies have been found in association with hydrocephalus. Thus hydrothorax, cleft-palate, web fingers and toes, club feet, absence of one kidney and many other abnormalities are reported in the literature. A consideration of the factors necessary for the production of hydrocephalus leads one to conclude that these are accidental findings and have no bearing on the development of the hydrocephalus.

Pathogenesis and Pathology.—In order to understand the manner in which internal hydrocephalus is produced it will be necessary to consider briefly the essential factors concerned, namely, the formation and the absorption of cerebrospinal fluid. Cerebrospinal fluid is formed within the ventricles by the activity of the choroid plexuses. Under normal conditions it passes from the ventricles through the foramen of Magendie and the foramina of Luschka directly to the spinal subarachnoid space and by way of the basal cisternæ to the cerebral subarachnoid space. From the basal cisternæ (cisterna cerebella medularis, cisterna pontis, cisterna interpeduncularis, cisterna chiasmatis) the cerebrospinal fluid is distributed along the major branches of the cerebral subarachnoid space which accompany the blood-vessels and traverse the sulci between the cerebral convolutions. Absorption in the ventricular system is negligible. From the subarachnoid system the cerebrospinal fluid is absorbed directly into the blood stream. Although absorption takes place from both the cerebral and the spinal subarachnoid systems, cerebral absorption is much greater than spinal absorption on account of the greater absorbing area. The normal balance between the formation and the absorption of cerebrospinal fluid depends on an adequate communication between the ventricular and the subarachnoid systems. When this is interfered with, hydrocephalus is produced. Interference with absorption of the cerebrospinal fluid, although there is adequate communication between the two systems also will bring about the retention of the cerebrospinal fluid and the development of hydrocephalus. Internal hydrocephalus may be due to one of these two factors, or, as is rarely the case, both factors may play a part in its production. It has been suggested that internal hydrocephalus may result from hypersecretion of the choroid plexus but the evidence for this is insufficient for it to be of more than theoretical interest.

The association of meningocele with internal hydrocephalus and the development of an internal hydrocephalus following the removal of a meningocele has been observed repeatedly. The combination of hydrocephalus and meningocele occurs so frequently that it is exceedingly difficult to say definitely that the removal of the meningocele caused the hydrocephalus. In many instances the hydrocephalus may not have reached a stage in its development which permitted of its recognition at the time of removal of the meningocele and that even without surgical interference the hydrocephalus would have become manifest at a later period. It has been suggested that a meningocele signifies an attempt on the part of the spinal membranes to compensate for the diminished absorption of the cerebrospinal fluid. If this were true the hydrocephalus would then develop because of the removal of the absorbing area of the meningocele, this having been just sufficient (or nearly so) to maintain the balance between the formation and the absorption of the cerebrospinal fluid. After its removal, the cerebrospinal fluid forms more rapidly than it is absorbed and consequently an accumulation of cerebrospinal fluid results. This explanation requires further substantiation before it can be accepted as the true cause of the hydrocephalus developing under such circumstances.

Two distinct types of internal hydrocephalus are recognized: *Obstructive* hydrocephalus and *Communicating* hydrocephalus. Obstructive hydrocephalus develops because the cerebrospinal fluid cannot escape from its place of origin in the ventricles to the cerebral and spinal subarachnoid space as the channels of communication between the two systems are occluded. In obstructive hydrocephalus the absorption of cerebrospinal fluid from the subarachnoid space is normal. Communicating hydrocephalus develops because the basal cisternæ, through which the cerebrospinal fluid passes before being distributed over the cerebral cortex, are obliterated, entirely or partly.

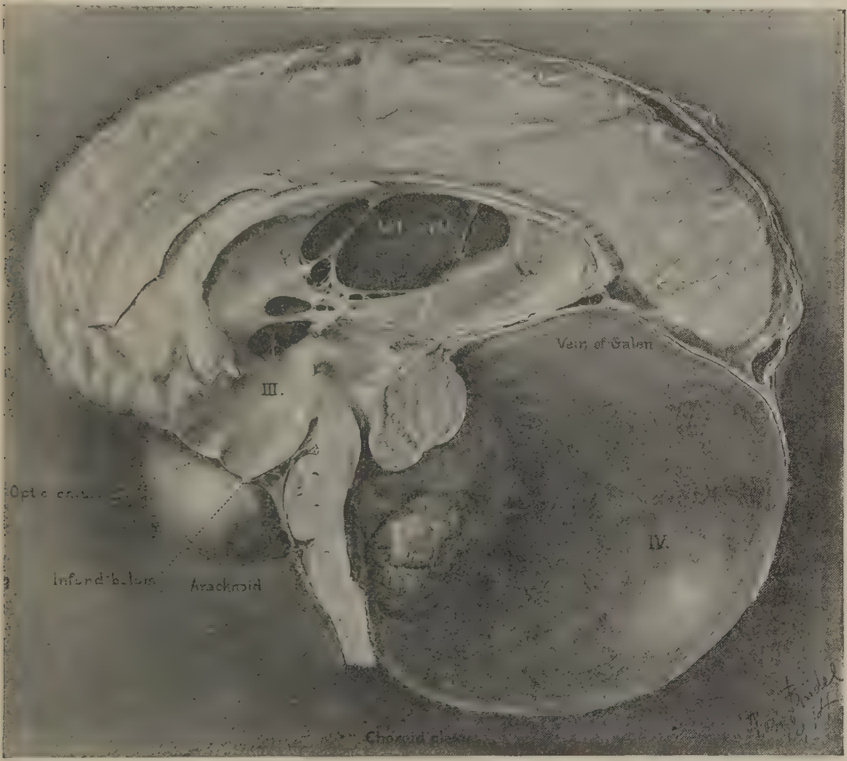

Fig. 55.—Midsagittal view of brain. The ventricles communicate freely with each other. The foramina of Magendie and Luschka have been occluded by adhesions. Note the enlarged foramen of Monro (arrow) and aqueduct of Sylvius. The large openings in the septum lucidum show the effects of the intraventricular pressure.

The channels of communication between the ventricles and the spinal subarachnoid space are patent to a greater or less degree.

The following resumé illustrates the different means by which hydrocephalus may be produced. If the outflow of fluid from one lateral ventricle is prevented by a tumor occluding the foramen of Monro a unilateral *obstructive* hydrocephalus results. If the aqueaqueduct of Sylvius is occluded or if the foramina at the base of the brain are obliterated an *obstructive* hydrocephalus takes place. On the

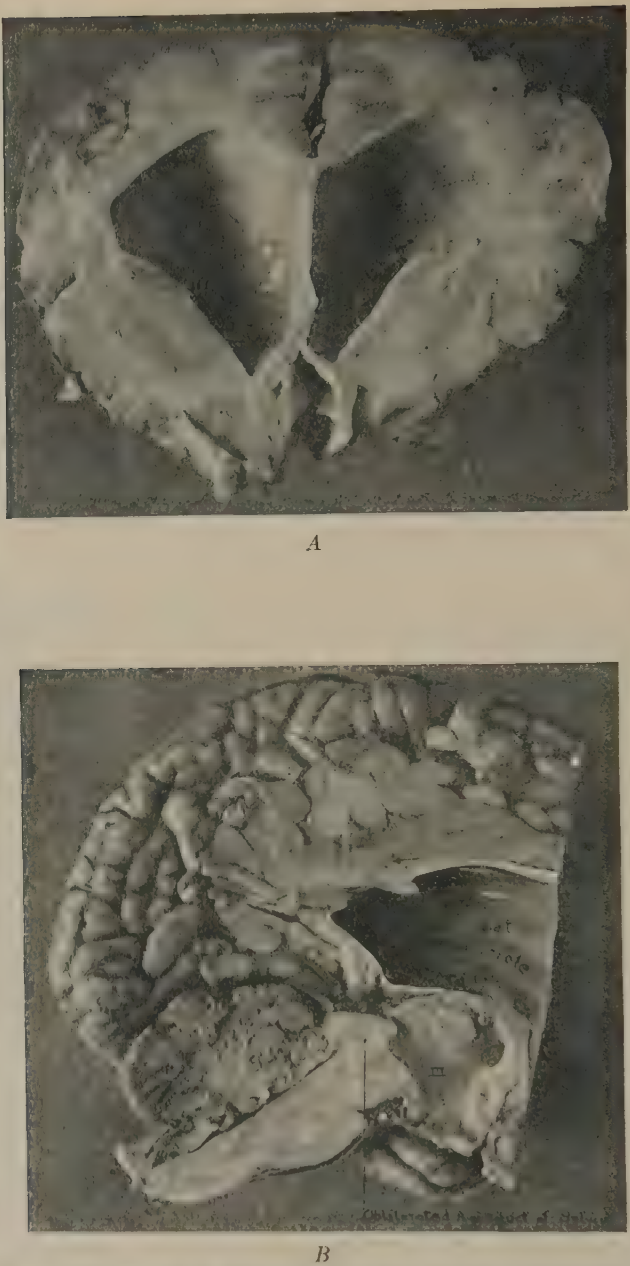

FIG. 56.—*A*, Cross-section of brain to show greatly distended lateral ventricles due to complete occlusion of the aqueduct of Sylvius. *B*, Midsagittal section of brain to show complete absence of the aqueduct of Sylvius and its replacement by neuroglia. Note the greatly dilated third and lateral ventricles anterior to the obstruction and the flattened slit-like fourth ventricle posterior to the obstruction. The septum lucidum is almost completely atrophied.

other hand, if the pathways of fluid from the ventricular system to the subarachnoid system are patent but if the basal cisternæ are more or less obliterated, the cerebrospinal fluid is prevented from being absorbed and a *communicating* hydrocephalus results.

In order to demonstrate the primary cause of the hydrocephalus, the brain and its coverings should be fixed *in situ* by injection of the cerebral blood-vessels with formalin. The brain should be removed with care in order to determine the patency or the occlusion of the foramina and the involvement of the basal cisternæ.

The pathologic changes in obstructive hydrocephalus depend upon the location of the obstruction. When it is at the base of the brain, the foramina of Magendie and Luschka are obliterated. This may be due to a tumor, to adhesions or an inflammatory exudate. Anterior to the obstruction, the fourth ventricle, the aqueduct of Sylvius, the foramina of Monro, the third ventricle and the lateral ventricles are greatly distended and filled with cerebrospinal fluid. The fourth ventricle may be so distended that it resembles a huge cyst (Fig. 55); in such an instance the cerebellar lobes are thinned, they are widely separated by the cyst which completely fills the posterior cranial

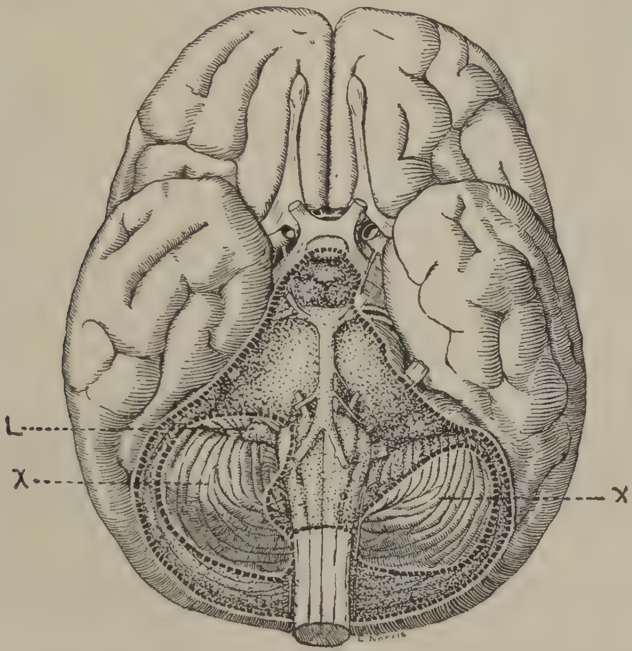

Fig. 57.—Diagram of the base of the brain, showing the distribution of the adhesions over the base of the brain responsible for the communicating type of internal hydrocephalus. *X*, *X* represent areas on the cerebellar hemispheres which are relatively free from adhesions. *L* represents the foramen of Luschka, which is patent only on this side, the opposite side is completely obliterated by adhesions. The adhesions extend along the tentorium on both the superior and inferior surfaces.

fossa. When the obstruction is anterior to the basal foramina in the region of the Aqueduct of Sylvius, the third and the lateral ventricles are distended with cerebrospinal fluid. The fourth ventricle is compressed and the cerebellar lobes are greatly flattened. Congenital absence of and pathologic changes in the aqueduct of Sylvius have been the causes of the obstruction in this location more often than any others in the cases which I have seen (Fig. 56). There usually is a small indentation where the aqueduct of Sylvius should begin in the third ventricle but beyond this it is not patent. The remains of the aqueduct of Sylvius may be demonstrated microscopically or it may have been replaced by a gliosis.

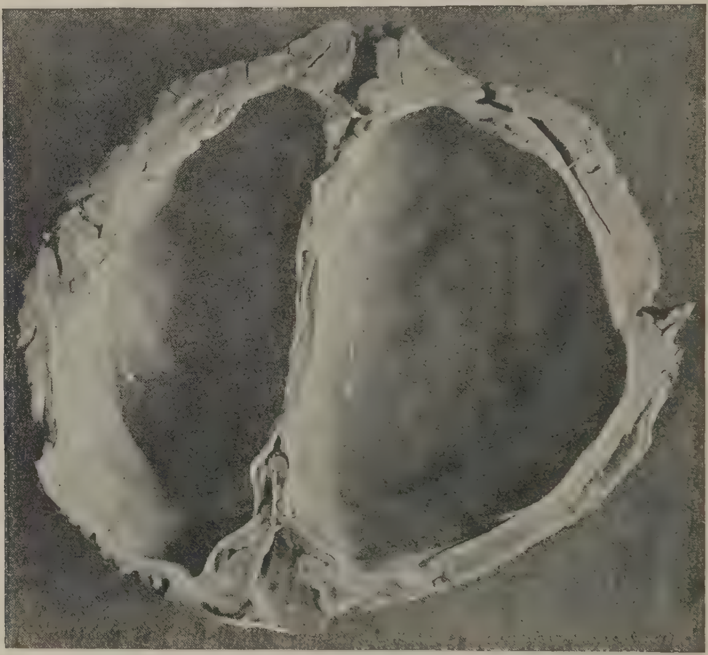

Fig. 58.—Communicating hydrocephalus. Cross-section of brain showing distended ventricles and thinning of the cerebral cortex.

The important lesions in communicating hydrocephalus are adhesions at the base of the brain, the result in many instances of a previous meningitis. The lesion is situated so that the normal function of the cisternæ and of the foramina is disturbed. The foramen of Magendie and one of the foramina of Luschka may be obliterated by adhesions while the other foramen of Luschka remains patent or both foramina of Luschka may be closed but the foramen of Magendie is open. At least, one of the foramina remains patent in this form of hydrocephalus. The basal cisternæ are entirely or in part obliterated by fibrous adhesions (Fig. 57). The adhesions are most dense, as a rule in the region of the medulla and the cerebellum, being less pronounced in the region of the chiasm. A few adhesions may be found over the surface of the brain. They may involve the base of the brain and dense bands of fibrous tissue are found binding the tentorium to the posterior surface of each occipital lobe and the superior surface of the cerebellum or they may encircle the mid-brain as it passes through the opening in the tentorium cerebelli. In the majority of instances the adhesions are not limited to the cerebrum but also involve the spina subarachnoid space. As described in obstructive hydrocephalus the foramina are dilated and the ventricles are distended with fluid.

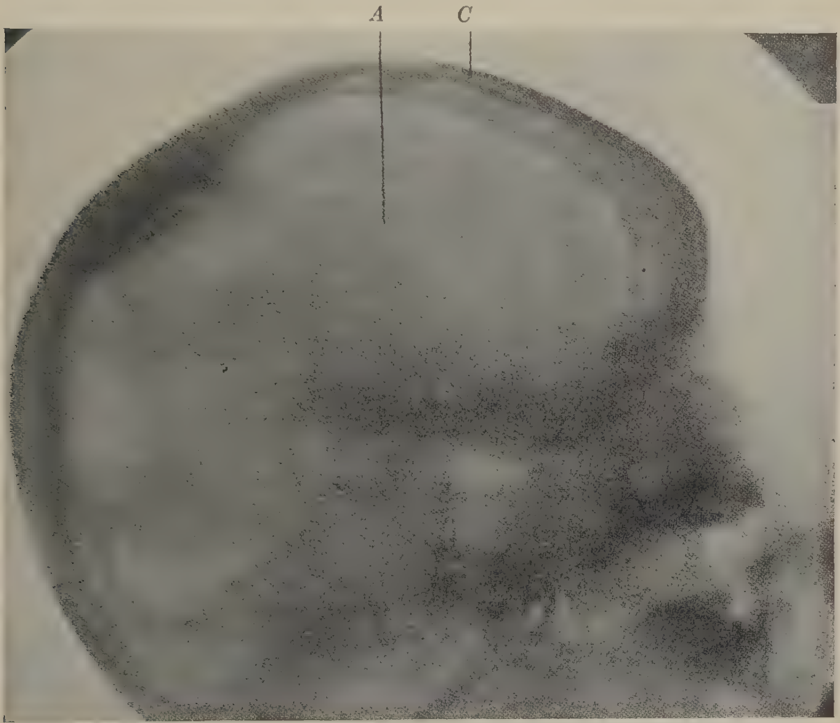

Fig. 59.—Roentgenogram taken after injection of air into ventricle. Note the ventricle filled with air (*A*). The cortex is of paper thinness (*C*). (Method devised by Dandy.)

The lesions in the brain and skull are essentially the same in both types of hydrocephalus and are dependent on its duration. The sulci are shallow, the convolutions are flattened and the cortex is so compressed that the gray and white substance is indistinguishable. The accumulation of cerebrospinal fluid distends the ventricles and there is a gradual compression and thinning of the brain substance (Fig. 58). If the condition exists over a long period of time only a thin film of cortex will remain (Fig. 59). The cortex may rupture and collapse into a shapeless mass. The septum lucidum becomes atrophied and completely disappears as the result of the continued intraventricular pressure or it may be represented by a few bands of tissue. The choroid plexus shows no characteristic pathologic changes.

The bones of the skull enlarge, become greatly distorted and thinned and areas of craniotabes due to atrophy of the bone develop. Wormian bones frequently form. The landmarks of the basal fossæ slowly become obliterated by the continuous pressure but as a rule the individual bones of the base of the skull do not become altered.

The cerebrospinal fluid contained within the ventricles varies in amount. Two liters and even greater amounts have been obtained from the ventricles or by lumbar puncture. The cerebrospinal fluid, whether obtained by ventricular or lumbar puncture usually is clear and contains from 6 to 12 lymphocytes per cubic millimeter. The globulin content is normal. In two cases of obstructive hydrocephalus the ventricular fluid was clear, yellow and contained red blood cells which indicated that there had been a recent hemorrhage into the ventricles. The cerebrospinal pressure while greatly increased in the communicating type is not increased in the obstructive type.

Symptoms.—The symptoms are similar in both types of hydrocephalus. Various stages occur. It should be remembered that the symptoms are due to the accumulation of cerebrospinal fluid within the ventricles, therefore the manifestations are dependent upon the rapidity with which the cerebrospinal fluid accumulates and the duration of the process. Closure of the fontanelle and union of the sutures influence the clinical picture of the disease. Congenital hydrocephalus is recognized "in utero" in a small percentage of cases: At times the process is so far advanced that delivery without operative interference is impossible. The "large head" may be noticed at birth but it is more common for the symptoms to attract no attention until a few weeks after birth when it is observed that the head is rapidly increasing in size. Acquired hydrocephalus most frequently follows a meningitis. The acute illness may subside and the patient appear to be recovering when attention is attracted by the progressive increase in the circumference of the head. In a small number of cases no history of a preceding illness can be obtained and the increase in the size of the head is the first evidence of the disease. The persistence of meningeal irritation or the return of the meningeal symptoms after they have subsided during the course of meningitis should always suggest the possibility of an hydrocephalus. The rapidity with which the symptoms develop is subject to wide variations. I have seen, in congenital hydrocephalus, the head increase 2.2 cm. in size over a period of four months, while, in another it increased 5.5 cm. within the same time. In a case of acquired hydrocephalus following meningococcus meningitis the head grew 7.5 cm. in seven months. As a rule the head enlarges more rapidly and reaches a greater size in congenital than in acquired hydrocephalus owing to the fact that the sutures are less firmly united. The head may enlarge slowly and be but little larger than normal in both the congenital and acquired forms. It is surprising what a large amount of cerebrospinal fluid may accumulate before there is any increase in the size of the head or definite cerebral symptoms become manifest. The rapidity of growth or the size of the head while no criterion as a rule of the seriousness of the condition become a matter of much importance in the treatment. When the process is fully developed, the shape of the head is so characteristic that it is not often mistaken for any other condition (Fig. 60). The head is enlarged out of all proportion to the face and body. In marked contrast to the enlarged head the nose, mouth and chin are small. One of the early and characteristic features of hydrocephalus is

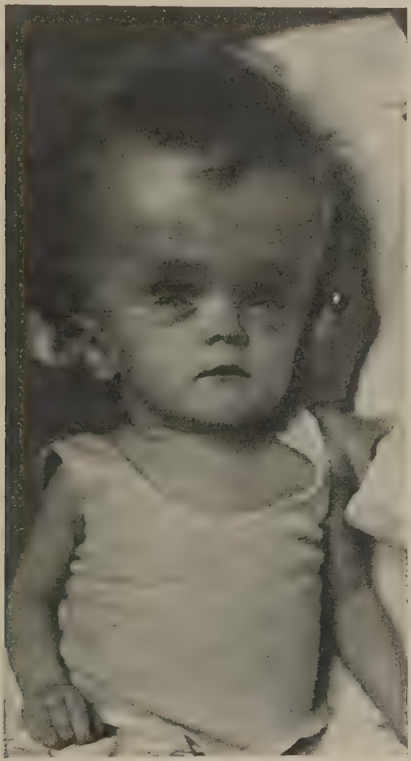

Fig. 60.—Communicating hydrocephalus (acquired). Note the head, enlarged out of all proportion to the face; the downward displacement of the eyeballs and the prominent frontal bones. The circumference of the head was 60 cm.

the displacement downward of the eye-balls so that the sclera which is normally hidden behind the upper eyelid is readily seen. This is due to the continued pressure on the orbital roof which is one of the earliest pathologic changes. The anterior and posterior fontanelles are open and bulging and the sutures are widely separated. There is marked prominence of the bones of the skull which in extreme cases amounts to an actual bulging of the frontal, parietal and occipital bones. Very early the veins of the scalp and the eyelids become prominently distended. The ears are small in contrast to the large head and the external auditory canal is pressed downward so that it may become a mere slit. The circumference of the head may reach 70 or 80 cm. and the weight of the fluid is such that the patient cannot raise his head or move it in any direction; as a consequence, decubitus ulcers are prone to occur. The increase in the intraventricular pressure is so gradual and so well compensated for by the compression of the cortex and the separation of the sutures that for a long time cerebral manifestations may not appear or vision be disturbed. Nystagmus and convergent strabismus are often present. Choked disc is rarely seen in chronic internal hydrocephalus but eventually optic atrophy results. Blindness may occur. As mentioned above cerebral symptoms are not prominent in the earlier stages but sooner or later the continued increase of the intracranial pressure causes hyperesthesia; ataxia and spasticity of the muscles of the extremities. The lower extremities are usually involved before the upper extremities. The reflexes are hyperactive. Convulsions are uncommon. Hydrocephalic children as a rule are quiet, inactive and well nourished. The cerebral manifestations are markedly increased when the patients are disturbed. The physical and mental development of the children is retarded. Intelligence is frequently so impaired that they may become idiotic. There is, however, great variation in this respect. I observed one child with an obstructive hydrocephalus who lived to be five years of age. He was never able to move his head and he was blind yet he understood simple commands, asked for food and recognized the voice of his parents. At post-mortem examination the cortex of the brain measured 1 cm. in thickness. The course of the disease is progressive unless the process becomes spontaneously arrested. When this occurs in the early stage the physical and mental development of the patient may not be affected. The disease is a chronic one and the patients may live for many months. I have known two patients, one of whom lived five years and the other seven years but this is the exception rather than the rule. Spontaneous rupture of the cerebral cortex may take place or death may occur from an intercurrent infection.

Diagnosis.—The characteristic enlargement of the head, the downward displacement of the eye-balls and the course of the disease in the advanced cases of hydrocephalus render the diagnosis comparatively easy. In the early stages of the disease the diagnosis is oftentimes difficult and can be established only by careful observation for a long period of time. It is of the utmost importance to establish the diagnosis as early as possible in order to relieve the primary cause of the hydrocephalus before the secondary changes have progressed to an advanced degree. Conditions most frequently confounded with hydrocephalus are rickets, pachymeningitis interna hemorrhagica and tumors. Rickets is often confused with hydrocephalus both on account of the large head and the tense and bulging fontanelle. The cerebrospinal fluid and pressure may be increased in each disease. The characteristic appearance of the frontal and parietal bones, the absence of the downward displacement of the eye-balls and the evidences of rickets elsewhere are signs which favor the diagnosis of rickets. In pachymeningitis interna hemorrhagica, bloody or yellowish tinged fluid is obtained from the subdural space and the fluid obtained from the ventricles generally is clear. The ventricles are not dilated. When the enlargement of the head is due to a tumor there may or may not be an associated hydrocephalus. The absence of focal symptoms favors the diagnosis of hydrocephalus. In chronic basilar meningitis an internal hydrocephalus is invariably present.

There is no way by which a clinical differentiation can be made between the obstructive and the communicating type of internal hydrocephalus except by the actual determination of the presence or the absence of communication between the ventricles and the subarachnoid space. The amount of cerebrospinal fluid obtained by lumbar puncture is not an absolute index as to the type of hydrocephalus unless as in advanced cases excessive amounts are withdrawn. Twenty-five cubic centimeters of fluid are frequently obtained when the obstruction is at the aqueduct of Sylvius as the secretion from the choroid plexus of the fourth ventricle is not disturbed. When the foramina at the base of the brain are completely obstructed, the amount of fluid is much less but even in such cases a small amount may be obtained.

The early diagnosis and the differentiation between the two types of hydrocephalus can be made with the *phenolsulphonephthalein test.* This test is based on a study of a large series of cases and gives accurate information as to the presence of an hydrocephalus and of the patency or occlusion of the foramina between the ventricular and the subarachnoid systems. By means of this test the absorption of cerebrospinal fluid from the subarachnoid space can be estimated. In obstructive hydrocephalus, phenolsulphonephthalein when injected into the lumbar subarachnoid space does not appear in the cerebrospinal fluid which is obtained from the ventricle within 40 minutes, if at all, while in communicating hydrocephalus it appears promptly. In communicating hydrocephalus the absorption of the dye from the subarachnoid space is greatly diminished, less than 20 per cent. being excreted in the urine in two hours as compared to 35 to 60 per cent. in obstructive hydrocephalus and in normal patients.*

1. To determine the degree of absorption from the subarachnoid space:

1 c.c. of phenolsulphonephthalein† with 1 c.c. of cerebrospinal fluid is injected into the lumbar subarachnoid space; the urine is collected for the period of two hours.

In normal patients 35+ to 60 per cent. is excreted in two hours.

In the communicating hydrocephalus less than 20 per cent. is excreted.

In the obstructive hydrocephalus 35 per cent. or more is excreted.

2. To determine the patency or the closure of the foramina between the ventricles and the subarachnoid space: After having injected phenolsulphonephthalein into the lumbar subarachnoid space, a ventricular puncture is made.

* The phenolsulphonephthalein test is carried out as follows:

† A freshly prepared, neutral solution of phenolsulphonephthalein should be used for these tests. This may be obtained from Hynson, Westcott and Dunning, Baltimore, Md.

If the foramina are patent, phenolsulphonephthalein appears in the ventricle within 1 to 20 minutes (communicating hydrocephalus). If there is an obstruction it does not appear within that time and may not appear at all (obstructive hydrocephalus).

3. A kidney function test should be made in the communicating type of hydrocephalus.

Prognosis.—Patients born with a rapidly developing hydrocephalus seldom survive infancy. When the hydrocephalus is due to a congenital obstruction of the aqueduct of Sylvius spontaneous recovery is impossible but when it is due to adhesions, it is not difficult to imagine recovery following their gradual disappearance. However, unless the primary cause of the disease can be ascertained and relieved, a satisfactory outcome should not be looked for. Retrogression of the symptoms is exceedingly uncommon although when it does take place the patient may not be mentally or physically handicapped.

Treatment.—Various operative procedures have been employed for the relief of chronic internal hydrocephalus. Puncturing the ventricle, the establishment of a communication between the ventricles and the cerebral subarachnoid spaces, drainage into the jugular vein and into the retroperitoneal space have given uniformly discouraging results and are rarely of permanent value. Drainage of cerebrospinal fluid from the ventricles by repeated lumbar punctures is a procedure of questionable value and cannot be recommended except as a temporary measure. The communicating type is the only one in which this method offers any benefit.

Dandy has devised recently two surgical methods based on the pathology of internal hydrocephalus. He has successfully reëstablished a communication between the ventricles and the subarachnoid space in obstructive hydrocephalus and he has diminished the formation of cerebrospinal fluid by removing the choroid plexus of the lateral ventricles in communicating hydrocephalus. These new procedures offer promise of being of value in the treatment of this disease.

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CHAPTER CLXXVI

THROMBOSIS OF THE CEREBRAL SINUSES

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Two forms of thrombosis of the cerebral sinuses are distinguished according to the underlying conditions with which they are associated. Primary or marantic thrombosis occurs during the course of long and exhaustive disease; secondary or infective thrombosis results from the extension of an inflammatory process from a primary focus of infection in the neighboring tissues. Rarely thrombosis follows a traumatic or operative injury.

The symptoms in primary sinus thrombosis are due to venous stasis and the consequent circulatory disturbances of the brain. In secondary thrombosis there may be in addition to the cerebral disturbances a general septicemia. Both forms of thrombosis are relatively less frequent in infants and young children than in adults.

Primary or marantic thrombosis occurs in association with marasmus, the severe gastro-intestinal diseases, pertussis, pneumonia, diphtheria and other acute infectious diseases. It may develop during the course of any disease in which there is a general systemic weakness. Too much importance has been given to the association of cerebral thrombosis with marasmus, the gastro-intestinal diseases and the acute infectious diseases in infants and young children. Holt did not find a single instance of cerebral thrombosis in over 200 autopsies in infants dying from nutritional disturbances. It is infrequent also in acute infectious diseases.

The longitudinal sinus is most frequently involved in primary thrombosis. The thrombus may extend to the other cranial sinuses and involve the cranial veins. It begins usually at the middle of the sinus or at the top of the cranial cavity; it may extend forward or toward the torcular Herophili. Involvement of the other sinuses always results from a direct extension of the process to them. The thrombus at first soft in consistence and moist, later by contraction and extraction of fluid, becomes compact, firm, dry and granular in texture. As a consequence of the thrombosis there is congestion of the cerebral vessels and minute hemorrhages may be found scattered over the cortex of the brain. Larger hemorrhages may result from rupture of the meningeal or cerebral vessels. When circulation is rëestablished, it is brought about through a rechanneling of the thrombus. The thrombus may be absorbed or become organized. Such an outcome is uncommon.

The symptoms may be masked by the manifestations of the primary disease but if the thrombus forms after the acute symptoms have subsided, the first evidences of its presence are headache, delirium, vomiting, convulsions (localized or general) and coma. The veins over the scalp and eyelids become dilated. The retinal vessels are distended and choked disc may appear early. Paralysis of the extremities may develop.

The diagnosis is made by the dilatation of the superficial veins of the eyelids and of the forehead and by edema of the scalp. Thrombosis of the retinal vessels may be of aid. The possibility of a sinus thrombosis should always be considered when convulsions and loss of consciousness develop during the course of any prolonged and exhaustive illness.

The course may be so short that the symptoms are regarded as terminal manifestations of the primary disease and the thrombus is "accidentally" found at the pathologic examination. The thrombosis in rare instances exists for a long time before death intervenes.

The treatment should be directed toward the cure of the primary disease and improvement in the general condition of the patient as there are no therapeutic measures which influence the thrombosis after it has formed. Surgical intervention should be considered; especially when the thrombosis develops after a traumatic or operative injury.

Secondary or Infective Thrombosis.—In this form the thrombosis results from the direct extension of the infection to the sinus wall or from a thrombus in the small veins to the sinus into which they empty. It occurs as a complication in erysipelas, furunculosis and other infections of the scalp or face. The most common causes of secondary thrombosis, however, are suppurative processes in the middle ear and the accessory sinuses of the nose. The reason for this is the anatomic relationship between the mastoid cells and the sigmoid sinuses and the proximity of the sphenoidal cavity to the cavernous sinuses. Acute and chronic infection of the middle ear constitute a large proportion of the conditions which are seen in children yet secondary thrombosis of the cerebral sinuses is comparatively uncommon. The infrequency of thrombosis in these conditions is accounted for by the anatomic peculiarities of the mastoid cells at this age. There is the opportunity for free drainage to be established more readily and for the suppurating process to appear externally rather than to extend to the sinus wall. The writer has seen only six cases in his experience.

Thrombosis of the Sigmoid Sinus.—A long standing suppuration of the mastoid cells may exist without resulting in thrombosis. If the process destroys the thin shell of bone separating the mastoid cells from the sigmoid sinus, the pus may extend to the external surface, or it may break through the dura and lead to a dural abscess or to meningitis. However, if there is an erosion of the bones and the inflammatory process involves the sinus wall, with destruction of the endothelium, a partial or complete thrombus will form. In the early stage the thrombus is adherent to the sinus wall, later suppuration often occurs and the sinus becomes filled with pus. The thrombus may become organized. The process in the sigmoid sinus may extend to the sagittal sinus and to the jugular vein. When the petrosal sinus is first affected, the thrombus may spread forward to the cavernous sinus or backward to involve the sigmoid sinus.

The general symptoms are those of septicemia. The irregular temperature with marked remissions and chills may be the only evidences of thrombosis aside from the local signs in the middle ear. Chills followed by profuse perspiration are common in adults, but in children the signs of collapse are more frequent (grayish pallor, cyanosis, cold hands and feet). Headache, general or local, nausea, vomiting and irritability are frequent symptoms. If the thrombus spreads and breaks down, infected particles are carried into the general circulation and metastatic abscesses form. Metastases occur most frequently in the lungs, in the liver and in the spleen. Abscesses locate less frequently in the joints, in the eye and in the brain. The liver and spleen become enlarged. Convulsions are rare and in contradistinction to other cerebral conditions, the intelligence of the patient is usually retained to the end. There is a polymorphonuclear leukocytosis. The spinal fluid is normal except when meningitis or cerebral abscess complicates the thrombosis. When this complication ensues the spinal fluid is under increased pressure, turbid and there is an increase in the cellular elements. Cultures from the spinal fluid may be sterile, but sooner or later organisms are found. Considerable attention has been given to the importance of blood cultures in sinus thrombosis and it is now generally believed that a bacteremia is present at some time or other in nearly all cases. The bacteremia may occur early. In many instances the organisms are not constantly present and are found only when cultures are made repeatedly. After operation with ligation of the jugular vein the organisms disappear from the blood rapidly; within 24 to 48 hours the cultures may be sterile. The streptococcus is the organism which is present most often. Staphylococcus aureus occurs next in frequency The pneumococcus is found rarely. Local manifestation of thrombosis map or may not be present. A localized and painful edema along the posterior border of the mastoid is suggestive of thrombosis. Care should be taken not to confuse this with the swelling and tenderness due to inflammation of the mastoid itself. Extension of the thrombus to the jugular vein map be detected by tenderness over its course. The thrombosed vessel may be palpated. The patient invariably holds his head turned to the affected side. Other evidences of thrombosis of the sigmoid sinus (hoarseness, dyspnea, slow pulse and spasm of the sternocleidomastoid muscle) may result from pressure on the vagus and the spinal accessory nerves as they pass through the jugular foramen with the thrombosed sinus. Eye changes, such as choked disc, nystagmus and strabismus may be present. Involvement of the sinus may be suspected by local evidences of middle-ear disease or by the general symptoms.

Thrombosis of the Cavernous Sinus.—This is usually the result of a primary suppurative condition in the sphenoidal cells with extension directly to the cavernous sinuses. Occasionally the thrombosis results from an infection in the region of the orbit with thrombosis of the ophthalmic veins and from them the thrombus extends to the cavernous sinus; it may extend from the sigmoid sinus. In addition to the symptoms of septicemia, there are the local manifestations of thrombosis of the cavernous sinus. They are definite and characteristic. There is marked exophthalmos, edema of the eyelids, choked disc and retinal hemorrhages. The veins in the region of the orbit are congested and the oculomotor nerves are affected so that the eyeball with its dilated pupil is immovable. The thrombus rarely is localized to one sinus but extends through the sinus of Ridley to the adjacent sinus. Therefore, first one sinus is involved and then the other or there may be bilateral involvement.

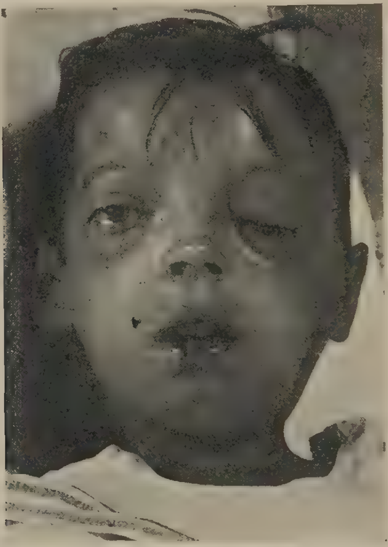

Fig. 61.—F. K., 12 years. The illness began three days after swimming and diving in a public swimming pool. The first symptoms were swelling and exophthalmos of the right eye. They later extended to the left eye. After three days the eye-balls became fixed and there was a bilateral choked disc. The temperature was of the septic type. Metastases appeared in the bones of the skull, the lungs and the left knee. The staphylococcus aureus was grown from the blood and the urine. The patient recovered after an illness lasting 3½ months.

The **diagnosis** of secondary or infective thrombosis is not difficult when there is a primary focus of infection, chills, a septic temperature and local manifestations of obstruction to the sinus. In suppurative conditions of the middle ear, thrombosis of the sigmoid sinus should always be suspected when there is a high temperature with remissions, chills, enlarged spleen and leukocytosis. In such cases a positive blood culture is of great help in establishing the diagnosis, even in the absence of definite symptoms of obstruction to the sinus. Exophthalmos, edema of the eyelids and choked disc are pathognomonic of cavernous sinus thrombosis.

The **prognosis** depends on the location of the thrombus, the severity of the infection and the complications. Although the average duration in untreated cases is from six to eight weeks, it may run a much longer course. An early diagnosis permitting of early operative treatment is most important in determining a favorable outcome. In cavernous sinus thrombosis death from meningitis or septicemia is almost inevitable. Instances of spontaneous recovery are rare (Fig. 61).

The **treatment** of thrombosis of the sigmoid sinus consists in evacuating the infectious process in the mastoid cells and antrum together with the opening of the sinus and the removal of the thrombus. It is generally advised to ligate the internal jugular vein. Operative measures for thrombosis of the cavernous sinus have been unsuccessful.

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CHAPTER CLXXVII

BRAIN PARASITES

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The only important brain parasites are the echinococcus and the cysticercus. Both are usually associated with a general ivasion of the body by the parasite. They are very rare in children. In 102 cases Sato found only three in the first decade of life and seven in the second.

The cysts in the brain are usually not large, but often multiple and bizarre in shape. They are often surrounded by an area of cerebral softening. The parasites may be in the ventricles and they may be floating about in the spinal fluid. In several instances the diagnosis has been made by finding the parasites in the cerebrospinal fluid obtained by lumbar puncture.

The **symptoms** are those of a small brain tumor. The general symptoms are often not marked—headache and choked disc may be absent, etc. The localizing signs are varied. Epileptic attacks may be the only indication of their presence. In the case reported by Cade there was an hemianopia. In some cases there are signs of meningeal irritation, so that a diagnosis of meningitis is thought of. Other cases are described as showing mental symptoms, especially mental confusion and agitation. It is not uncommon for the symptoms to subside temporarily or even for a year or more. The invasion of the fourth ventricle may be the cause of sudden death.

The **diagnosis** cannot be made positively unless the patient is known to be infected elsewhere, liver, etc., and the cerebral symptoms show the brain to be involved. Examination of the spinal fluid or a diagnostic brain puncture may show the parasite. In many cases the condition is found at necropsy when not suspected.

Treatment is operative. The cysticercus is more frequently multiple than the echinococcus and is therefore less likely curable. The literature shows that few patients survive the operation.

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CHAPTER CLXXVIII

ABSCESS OF THE BRAIN

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An abscess of the brain is much like that seen in other tissues and like them is due to the presence of a microörganism and a collection of pus more or less walled off from surrounding healthy tissue by a fibrinous or fibrous wall. The etiology of brain abscess may be referred to anything that causes the deposition of bacteria in a focus favorable for their multiplication and the demonstration of their locally irritant qualities. In the case of brain abscess this is most frequently by way of the blood stream an infected embolus from a gangrenous lung or other focus elsewhere in the body. The development of a brain abscess also follows blows on the head which are not severe and show only the symptoms of a mild concussion of the brain and without in any way giving a means of egress of microörganisms. The reason for this is that the concussion produces a lowered resistance in the brain tissue to pathogenic organisms already in the circulation and this spot of lowered resistance is rapidly seized upon as a suitable medium for their growth. Brain abscess may also be caused by the direct extension of infection from infected foci in the neighborhood: Frontal sinus disease, osteitis of the bones of the skull, panophthalmitis, and most frequently of all, from middle ear disease or mastoid sinus infection. Another cause of brain abscess is the penetration of the brain by infected missiles.

Symptoms.—The first stage in the development of a brain abscess is an acute but more or less localized encephalitis. This is sometimes spoken of as the initial stage. There is usually some headache but the more marked symptoms are the general reaction: Chill or sensations of chilliness, fever, rise in pulse rate, leukocytosis and feeling of general malaise. There may also be symptoms indicative of the irritation of some part of the brain such as Jacksonian epileptic attacks if the lesion is near the motor area, etc. With the formation of the abscess wall the general symptoms are usually diminished in intensity and if a firm wall is formed the abscess may remain "latent" for a considerable period. Cases are on record in which an abscess has remained latent for 20 years and I have observed a case in which an abscess was found in the brain at necropsy on a patient that had died of another affection and with no symptoms of the abscess. After a variable length of time the abscess enlarges and in so doing causes symptoms like those of a tumor of the brain. There are the same general symptoms of increased intracranial pressure:

Headache, projectile vomiting, slow pulse, disturbance of consciousness, epileptiform attacks and choking of the optic discs. In addition there are the "localizing" signs which are the same as those caused by tumor in the same locations. In this connection it should be borne in mind that the favorite sites for brain abscess are the frontal lobes, the temporosphenoidal lobes and the cerebellar lobes. In the case of the first two the localizing symptoms may be absent, as these are the so-called silent areas of the brain. In either of these locations, however, it is only a question of time until the abscess enlarges sufficiently to impinge on the motor area and produce either localized epileptic attacks or partial hemiplegia. Abscess in the cerebellum may be recognized by the distinctive disturbance of equilibrium and the ataxia that is caused especially the hemi-asynergy, adiadiokokinesis, etc. and, furthermore they are almost always the result of extension of infection from the mastoid or an infected sinus thrombosis.

In its further course an abscess may: First, cease to enlarge, become thickly walled and give no further symptoms; it may continue to enlarge and cause death either from the increasing pressure or through a rupture into the ventricle or into the meninges causing a meningitis; or it may be opened by the surgeon and its contents evacuated.

The **prognosis** of brain abscess is not invariably bad. Some patients recover spontaneously. The results of operation depend so largely upon the location of the abscess, the general condition of the patient, the accuracy of the localizing diagnosis and especially upon the skill of the operator that no general figures can be given. In my personal observation of over 20 cases, operated by several different surgeons, only one case had a fatal termination and in that one the result was due to the delay in diagnosis.

The **diagnosis** of brain abscess from meningitis may be extremely difficult. In fact meningitis is not an infrequent complication. The spinal fluid findings in brain abscess may be negative but if the abscess is located near the meninges as is common, the meningeal reaction will account for some increase in the albumen and an increased cell count. Usually the rigidity of the neck and Kernig's and Brudzinzki's sign are absent in brain abscess but not always. In abscess there are more signs of increased intracranial pressure, such as choked disc, etc. and also more localizing signs such as aphasias, hemiplegia and so on. A brain tumor case may resemble a brain abscess especially if the tumor signs develop rapidly and if the patient has some coincident infection. A patient under my observation had a double mastoid infection which was operated. A week later she became drowsy and developed a right hemiplegia with aphasia. There were choked discs and projectile vomiting. Operation disclosed no abscess but a markedly dilated lateral ventricle. The ventricle was tapped with relief of all symptoms for a short time. Later they returned and at necropsy there was found a small tumor—glioma near the foramen of Munro which had blocked this foramen and caused the unilateral hydrocephalus. Acute encephalitis may cause some of the symptoms of brain abscess—in fact it may be the early stage of an abscess but the more marked symptoms of increased intracranial pressure are usually absent. Acute brain swelling, pseudotumor cerebri, may cause difficulty in diagnosis especially if there is a history of traumatism as there often is in such cases but there is not the fever and leukocytosis that is present in most abscess cases. Infected sinus thrombosis is also a cause of error but the localizing signs in such cases—if there are any—will show that it is either the region of the carotid sinus or jugular that is affected and both would be rare sites for abscess.

The **treatment** of brain abscess is mainly surgical. In the initial stage an operation is not indicated except to remove the primary focus of infection if such be found. The patient should be absolutely quiet and with an ice-bag to the head. Antiseptics, such as urotropin, may be of some value. As soon as the condition is diagnosed as a definite abscess an operation should be done but from personal observation it seems to me that certain points in the technique should be carefully observed. First, the operation should be in two stages. In the first stage the overlying bone should be removed and the dura opened, irritated slightly and then replaced. The object of this is to produce adhesions between the dura and pia all around the spot where the abscess will be opened. In 24 to 36 hours the skin flap is again reflected and the abscess incised freely—not simply aspirated. The opening should be large enough so that the operator can see the entire abscess wall and carefully swab it out. It is then packed lightly. In 24 hours or less it is again swabbed out, using a speculum so that all parts are seen. Dressing must be continued daily until the abscess is healed. The advantage of the two stage operation is not only to produce adhesions but also to coax the abscess to point near the opening desired so that the least possible amount of brain tissue must be incised to reach it. If it is not freely opened and packed the walls collapse irregularly leaving pockets of pus which cannot be thoroughly cleaned out. If this happens the symptoms are relieved for the time but there is great danger of the formation of multiple new abscesses.

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CHAPTER CLXXIX

TUMORS OF THE BRAIN

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Tumor of the brain may occur at any age and its incidence is apparently not influenced by climate, race or similar factors. Its frequency in childhood is about the same as in adults.

The **etiology** of cerebral neoplasm, in most cases, cannot be determined. In childhood teratoid growths, congenital cysts and hemangiomata may be expected. They usually occur in the vicinity of the median line and may show no symptoms during early childhood. I have personally known of several instances in which there were no symptoms until the age of 10 or 12 years, although the autopsy showed beyond doubt, that their existence was congenital. Sometimes these formations are found only accidentally, at a post-mortem and have caused no symptoms during life.

Traumatism is frequently given as the cause of the development of brain tumor. In childhood where falls are frequent and the relation between cause and effect not especially clear, it is quite common to obtain a history that all of the symptoms developed shortly after a fall in which the head was struck. In most cases this apparent relation is misleading. There is little question, however, but that in some cases there is a real relation between the two. In some of these the growth has existed before, but has been stimulated by the traumatism. This must be the case where serious symptoms develop soon after the trauma and are followed by death and in which the autopsy shows a large tumor. The introduction of the foreign body into the brain, as for instance the spicule of bone from a depressed fracture may be the starting point of a tumor. A case of this kind is recorded by Leszynsky.* Dr. D. J. McCarthy has made some experiments on animals which confirm this possibility. It seems quite possible that brain tumor may develop in the cicatrix from an old encephalitis, either an infectious or traumatic encephalitis. The relation of trauma to brain tumor is a much debated subject and has been thoroughly reviewed by Hubschmann.† Interesting cases of this sort are reported by Strauss.‡

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* Jour. of the Amer. Med. Assoc. (1907).

† Deut. Zeitschr. F. Nervenheilk., lxvi, 1920 and also in the Revue Neurologique, 1920, No. 2, p. 163.

‡ Trans. of the Amer. Neur. Assoc. (1918), p. 114.

The infectious granulomata: Tuberculoma and gumma, especially the former, are not infrequently found in children. The solitary tubercle of the brain may exist in the absence of obvious tuberculous infection elsewhere in the body; the point of entrance of the tubercle bacilli, in these cases, remaining obscure. Syphiloma may be the result of inherited syphilis or the result of an infection from a nurse or by being kissed by a person with the disease. In this connection, it should be remembered that children who have inherited syphilis not infrequently develop anomalies and teratoid formations. I saw a case of this in a boy, age nine years, who presented numerous stigmata of hereditary syphilis and had a strongly positive Wassermann reaction on his blood. He had symptoms indicating a brain tumor and a gumma was thought of. He grew steadily worse in spite of antiluetic treatment and at necropsy it was found that although he had syphilis of practically all the viscera yet the central nervous system was free from syphilitic involvement. He had a large teratoid growth at the base of the brain.

Carcinoma of the brain is rare in childhood. It is practically always metastatic. I have seen a case of hypernephroma, in a boy of 12, in which there was a metastatic tumor of the brain.

Parasitic infections, such as the echinococcus, may be carried to the brain and develop characteristic cysts and give rise to symptoms of tumor of the brain.

Pathology.—The variety of tumors which may be present within the cranial cavity, are as numerous as may develop elsewhere. In children perhaps the most frequent are the tuberculomata and the gliomata (Dana and others). In my personal experience the tuberculoma although perhaps more common in children than in adults, is a rare tumor. The most frequent have been teratoid growths, gliomata and sarcomata. However, statistics of the relative frequency of brain tumors, in general, are of small value in determining the kind of tumor present in the individual case that is being considered.

The location of the brain tumor has a general relation to its histologic type. For instance, the solitary tubercle is almost always found in the pons or cerebellum. The sarcoma or fibrosarcoma is a frequent tumor in the region of the cerebellar-pontile angle. Gliomata are almost always subcortical and arise frequently as tumors from the floor of the ventricles. The endotheliomata are most commonly found near the meninges. The metastatic carcinomata and occasionally also the sarcomata spread through the meninges producing symptoms and signs more suggestive of a chronic meningitis than of a brain tumor.

The rate of growth of brain tumors varies with the nature of the tumor and also varies from time to time frequently without apparent reason. It has been observed that following decompression operations a tumor may appear to grow much more rapidly than before.

From a practical standpoint it is important to consider not only the histology of the tumor, but also the pathologic effects of the tumor upon the surrounding tissue. Some of the brain tumors such as the sarcomata and the endotheliomata are usually encapsulated, others such as the gliomata are not usually encapsulated. In either event there is often found in the vicinity of the tumor areas of inflammatory reaction constituting an encephalitis and the intensity of this reaction often accounts for the marked variation in the severity of the symptoms. Another complication of the tumor is the presence of hemorrhage either within the tumor or in the neighborhood; this is especially the case where the tumor erodes a large blood-vessel.

Tumors sometimes exert effects quite disproportionate to their size, by reason of the fact that their location obstructs the flow of the cerebrospinal fluid. In a case under my own observation a small tumor, about the size of a hickory nut, obstructed the foramen of Monro on one side. The result was an enormous distension of the posterior horn of the lateral ventricle on that side, with the symptoms of a tumor in the occipital lobe. Tumors in the region of the aqueduct of Sylvius, such as tumors of the pineal body and the corpora quadrigemina and also tumors obstructing the fourth ventricle, cause internal hydrocephalus and dilation of the third and the lateral ventricles. The dilation of the third ventricle is not infrequently accompanied by dilation of the infundibulum and this in turn compresses both the pituitary gland and also the optic tracts. This causes misleading symptoms of tumor of the pituitary.

The increase in intracranial pressure causes a flattening of the cerebral convolutions and also frequently causes erosion of the skull bones by the convolutions, so that their outline is visible by *x*-ray examination. Tumors in almost any location of the brain, but especially those at the base of the brain, are likely to produce alterations in the configuration of the sella turcica and other changes in the bones at the base of the skull. A dilatation of the internal auditory meatus, visible on *x*-ray examination may be caused by tumor in the posterior fossæ of the skull.

In any study of the effects of brain tumor on brain function it should be remembered that the area destroyed by a tumor, or affected by its surrounding encephalitis, is probably in close functional relation with other parts of the brain which may be anatomically quite distant. Secondary degeneration in the fiber tracts connecting these parts will occur and there is often a secondary atrophy of these functionally related parts. This is often seen in cases of tumor of the frontal lobe in which there is a secondary atrophy of the opposite cerebellar lobe, etc.

Symptoms.—The symptoms of tumor of the brain may be divided into two quite distinct classes, the first class including those which indicate a general increase of intracranial pressure; the second class include those symptoms which show disturbance of function of some particular part of the brain. This second class, the localizing symptoms or signs are of great importance in event that a surgical removal of the tumor is contemplated. One of the greatest difficulties in the operative treatment is the accurate localization of the growth.

The general symptoms are:

(*a*) *Headaches.*—Headache is one of the most constant symptoms of tumor of the brain, although in rare cases it may be entirely absent. Its character varies. Most often it is of a dull throbbing type and quite general. More frequently than not it comes in attacks resembling migraine. In some cases it is present only at certain times i.e., when the stomach is empty or just after arising in the morning or when the head is placed in certain positions, etc. The intensity of the headache is not necessarily proportionate to the size of the tumor. I have observed in a number of cases that the headaches were relatively less severe in an advanced stage, than in an early stage of brain tumor.

(*b*) *Nausea and Vomiting.*—This symptom not infrequently accompanies the headaches and increases the resemblance to migraine however, it can occur quite independently of the headaches. In children especially this symptom may be misleading in attracting attention to the stomach as the cause of the trouble. Vomiting of the projectile type, i.e., in which the vomitus is thrown forcibly from the mouth, often without warning, is quite characteristic when present. The vomiting usually has no relation to food, but may be brought on by certain activities, such as bending over, or may be present only on arising in the morning.

(*c*) *Dizziness, Vertigo.*—Attacks of dizziness or vertigo are not uncommon in some stages of the disease. Usually the true vertigo is found only in cases of tumor of the posterior fossa, in other cases the dizziness is of various irregular types.

(*d*) *Epileptiform Attacks.*—Generalized epileptiform attacks may be the result of a tumor in almost any location in the brain. In most cases the attacks have no distinguishable differences from the attacks of so-called idiopathic epilepsy. Epileptiform seizures of peculiar type such as Jacksonian epilepsy, in which only one part or one side of the body is affected; cerebellar fits, characterized by posturizing and cerebellar catalepsy; the uncinate group, characterized by disturbance of taste and smell, all have a localizing value. Cases have been reported in which epilepsy has been for many years (10 to 30), the only sign of a tumor of the brain (Oppenheim, Bruns, Creite, Simmonds and others). This is more frequently found in cases of cystic congenital growths, hemangiomata, etc. In some cases the attacks are more hysteroidal in character. I have observed such attacks more often in tumors of the pituitary region.

(*e*) *Effects on Pulse, Respiration, etc.*—These symptoms are frequently present in the advanced stage or even in an early stage of tumor of the posterior fossa. The pulse rate is usually slowed, the respiration rate may be slow or may show a Cheyne-Stokes type or a Biot type, the latter being characterized by long pauses. The temperature is usually unaffected, but may be subnormal in some cases. Just before death in brain tumors there is frequently a rapid rise in temperature.

(*f*) *Choked Disc.*—Choked disc is a typical sign of increased intracranial pressure and sometimes reaches a height of seven or eight diopters. It may, however, be entirely absent throughout the course of the disease. A choked disc of over two diopters is strongly suggestive of brain tumor. Other conditions causing choked disc, with rare exceptions, do not cause a swelling of the same degree as a brain tumor. It is not uncommon that a choked disc develops suddenly in the course of a brain tumor, or to have the disc suddenly increase in swelling. The cause of these sudden developments may be ascribed to hemorrhage within the tumor or to some similar reason for a rapid increase in pressure. After a length of time the discs are likely to recede and a condition of secondary optic atrophy arises. A high degree of choked disc is not incompatible with perfect vision, but if it lasts any considerable length of time, blindness will be the result. This may occur even though the tumor has been removed in the meantime or a decompression operation has been done.

The cause of choked disc in brain tumor is probably a mechanical one. It, however, has no special relation to the actual intracranial pressure, but is rather the expression of the rate of increase of intracranial pressure. Consequently we often see a choked disc receding in spite of the growing tumor, but in these cases we assume that the rate of growth is lessening. A choked disc is often unequal in the two eyes, but the side with the greater swelling is not necessarily the side of the tumor. As the research of Dr. Walter Parker has shown, these inequalities are due to the differences in intra-ocular tension, the protrusion of the optic nerve being proportionate to the difference between the intracranial pressure and the intra-ocular tension.

(*g*) *Disturbance in consciousness* of various kinds may be found in patients with brain tumors, but are not invariable and a practically normal mental state is not incompatible with the diagnosis. The most common disturbance is a slight stupor or stupidity. Occasionally other mental disturbances occur, hallucinatory experiences etc., leading to the diagnosis of a psychosis. Attacks of coma may occur which need not necessarily be fatal, though they are more frequent and more severe near the termination of the case.

(*h*) *Enlargement of the head* may be a symptom of increased intracranial pressure in childhood. It does not occur in adults. Close examination of the head in such cases usually shows a secondary hydrocephalus due to obstruction to the flow of the spinal fluid.

Localizing Symptoms.—Tumors of the prefrontal lobe, i.e., that portion of the front lobe anterior to the precentral convolution may not manifest their presence by any disturbance in cerebral function especially if they are on the right side of the brain in a right-handed person. In the left frontal lobe they may cause mental symptoms, chiefly, in the nature of defects in judgement, tendency to senseless joking, euphoria, etc. Their presence is often first recognized by their extension backward to the Rolandic area. Tumors in this location are frequently unaccompanied by choked disc. A retrobulbar neuritis is said to be present in these cases (Foster Kennedy and others).

Rolandic Area, Precentral Convolution.—Cortical tumors in this region are likely to manifest their presence by Jacksonian epileptic attacks, the twitchings are at first limited to one side of the body, one arm, one side of the face, etc., later they become more extensive, involving an entire side or even becoming generalized. It is not infrequent to find that after an attack the part affected is weak for a time and after subsequent attacks this weakness increases as the tumor grows and destroys the cortical area. There is at first usually a monoplegia, which may finally become a hemiplegia. Attacks of transient weakness may occur instead of epileptic attacks, the weakness becoming more pronounced and of longer duration, as the case progresses. A gradually developing hemiplegia should always arouse a suspicion of brain tumor as a cause. A tumor at the upper portion of the precentral convolution, near the paracentral lobule, may exert pressure also on the opposite hemisphere also and cause a paraplegia.

It should be remembered that the symptoms of tumor in the motor zone may be caused by tumors which began in an adjacent, relatively "silent" area and only caused distinct symptoms when they impinge on the precentral convolution. Such a state of affairs may be suspected when the patient has had general symptoms of brain tumor for a long time but has only recently shown signs of motor irritation or paralysis. Tumors in the motor area are likely to manifest their presence early and are therefore among those which are most frequently operated upon with success. Tumors in the vicinity of the third frontal convolution on the left side will produce a motor aphasia in addition to the paralysis.

Parietal Region.—Tumors in this location are characterized chiefly by sensory disturbances in the opposite half of the body. They may be shown by "sensory epilepsy" i.e., attacks of numbness and parasthesia sometimes followed by jerking. The most important localizing sign is the presence of astereognosis, i.e., the inability to recognize an object when it is placed in the hand, the patient's eyes being closed. This sign may be present in spite of the retention of pain and tactile sensibility in the extremity.

Temporal Sphenoidal Lobe.—A tumor of the temporal sphenoidal on the right side may grow to considerable size without producing localizing symptoms. When on the left side in right handed people it is likely to produce the so-called auditory aphasia and various forms of paraphasia. In the growth of these tumors their inward extension leads to the involvement of the lenticular nucleus which causes disturbance in the emotional sphere, forced laughing or crying and sometimes a rhythmical tremor. The extension into the uncinate region causes disturbances of sense of smell or taste usually complained of as hallucinations of unpleasant odors which are sometimes followed by epileptiform seizures (the uncinate group of fits—Mills).

The Region of Corpus Callosum.—Tumors in this location are relatively rare and their localizing diagnosis is very difficult, perhaps the most characteristic symptom is the presence of apraxia, an inability to use objects of common use, although their nature and name is well known. The symptoms may suggest a simple type of progressive dementia, where the general symptoms of brain tumor are slight or absent.

Tumors in the Basal Ganglia.—Tumors in the region of the basal ganglia are usually first recognized by their effect on the internal capsule which is to produce a progressive hemiplegia. Occasionally it may be recognized that they are producing lenticular disturbances i.e., rhythmic tremor and disturbances of emotional expression. Tumors in the optic thalmus cause the thalamic syndrome described by Roussey, hemianesthesia, hemi-anopsia and a severe pain of central origin in the anesthetic side.

Occipital Lobe.—The location of tumors of the occipital lobe is recognized because they usually cause a hemi-anopsia. In the early stages the hemi-anopsia may be incomplete, consisting only of the involvement of a sector of the visual field or a disturbance of the color fields. If on the left side of a right-handed person there may be the alexia, i.e., an inability to understand written or printed language, although vision may be normal. It is as if the words were in some unknown language.

Tumors of the Corpora Quadrigemina.—Tumors of the quadrigemina betray their location chiefly by their effects in producing paralysis of the third cranial nerve which is in the floor of the aqueduct of Sylvius just beneath them, also they cause obstruction to the aqueduct and a secondary internal hydrocephalus. They may also cause a homonymous lateral hemi-anopsia from pressure on the optic tracts and external geniculate, if the tumor grows more to one side than the other. The involvement of the pineal gland is not uncommon. If the pressure of the tumor extends downwards, the oculomotor palsy may be accompanied by a hemiplegia (also partial hemi-anesthesia) of the opposite side of the body, Benedict's syndrome.

Tumors in the Floor of the Fourth Ventricle.—Pontile tumors in this location are not infrequent in children. They usually consist of gliomata which arise from the neurolgia of the floor of the ventricle or of solitary tubercles. Their chief localizing sign is the presence of crossed hemiplegia i.e., paralysis of one side of the face or tongue and the opposite side of the body, etc. They not infrequently extend up the roof of the ventricle or through that and into the cerebellum, producing various cerebellar symptoms such as incoördination, ataxia, etc. Choked disc is usually early and severe.

Medulla.—Tumor of the medulla is distinctly uncommon although not unknown. Large tumors would probably be theoretically incompatable with life, but I have seen a case in which the tumor attained considerable size. The localizing signs are likely to be a crossed hemiplegia, especially the syndrome of Smith or Avellis, a unilateral paralysis of the tongue or of the larynx with hemiplegia of the opposite side.

Cerebellar.—Tumors of the cerebellum may attain considerable size without distinguishing symptoms or signs especially if they are cortical and do not affect the intracerebellar nuclei. In general it may be said that tumor of the cerebellum produces disturbance in coördination, staggering gait, etc. Choked disc is likely to be an early symptom due to a secondary hydrocephalus. The symptoms are more marked upon the side of the tumor. Special signs of cerebellar involvement are described: Adiadiococinesis, the inability to stop a movement quickly and initiate the opposite movement which is usually tested by having the patient rapidly pronate and supinate the hand; hemi-asynergy, the lack on one side of the body of synergistic control of the muscles in performing purposeful acts such as striking or throwing a ball; dysmetria, a disturbance in the control of the force used in performing a movement, usually tested by having the patient touch a particular point when it will be noticed that he strikes the point unnecessarily hard or by having him draw a line between two points and it may be noticed that he stops short or overruns his objective. These tests are not conclusive in as much as they may be absent even though there is extensive destruction of one lobe of the cerebellum. A peculiar attitude of the body, inclination of the head to one side, or a disturbance of equilibrium, noticed when the patient is bending forward are also characteristic phenomena of cerebellar lesions. Dana described a peculiar type of attacks as occurring in disease of the cerebellum in which the patient assumes certain postures. I have repeatedly seen this sort of attack and almost invariably associated with disease of the cerebellum although in one case it occurred in connection with a cyst at the base of the brain (involvement of the cerebellar peduncle probably), and in another case it was associated with an external cerebellar cyst. A spontaneous nystagmus is not infrequently associated with cerebellar disease and the nystagmus and pointing tests of Barany are useful in the diagnosis, at least theoretically.

Tumors of the Cerebellar-pontile Angle.—This term is restricted to those tumors which are external to the brain stem although frequently compressing it and which lie in the angle formed by the cerebellum, medulla and pons. They are most frequently fibromata or fibo-sarcomata growing from the sheath of the facial or the acoustic nerve. They are usually of slow development, well encapsulated and removable by operation. The earliest symptom is likely to be a progressive deafness and a facial palsy of the peripheral type on the same side as the tumor. At the later stage these tumors, through compression of the pons, may produce a hemiplegia on the opposite side, or by compression of the cerebellum, some disturbance in coördination. They are not infrequently multiple, i.e., on both sides.

Tumors of the Pituitary Region.—Tumors of the pituitary gland itself are usually of adenoma or adeno-carcinomatous type. They may develop from the remnant of the pharyngeal duct. It is not infrequent to have other tumors of the base of the brain such as sarcomata, etc., grow down into the sella turcica and cause pituitary symptoms. The localizing diagnosis of the pituitary tumor depends upon the development of three groups of symptoms: (*a*) The signs of intracranial pressure as seen in the case of any brain tumor. In the case of pituitary tumors proper, these signs are not likely to be marked. The headache is of the migraine type and choked disc is usually absent, the optic nerve head showing primary optic atrophy instead. (*b*) Symptoms which are the result of involvement of the brain in the neighborhood of the tumor. These symptoms are likely to be marked only when the tumor is of considerable size and consist chiefly in disturbance of consciousness, sometimes partial hemiplegia, etc. The pressure of the tumor on the optic tracts leads to the development of a bitemporal hemi-anopsia, but if the tumor extends more to one side than the other this may result in the destruction of the optic tract on that side and blindness of the eye on the same side. (*c*) Symptoms referable to the internal secretion of the pituitary and in cases of hypopituitarism, the development of Fröhlich's dystrophia adiposo-genitalis syndrome. In some cases hyperactivity of the gland may result in acromegalic symptoms or gigantism and this may be followed by its destruction so that the one syndrome develops after the other. In a case under my observation this occurred as the result of an operation in which a portion of a sarcoma was removed.

The pineal gland tumor is also characterized by disturbances of growth and nutrition due to disturbance in the internal secretion. The pressure of such tumors upon the corpora quadrigemina leads to disturbances of vision and palsies of the extra-ocular muscles.

Diagnosis.—The diagnosis of tumor of the brain depends upon the history and the presence of general signs of increased intracranial pressure together with some definite evidence of a localized disturbance of brain function, i.e., localizing signs. In the absence of either of these factors, a diagnosis must be considered doubtful.

The conditions which simulate tumor of the brain are: Brain abscess; the symptoms are all of the same type and character as of brain tumor, but are likely to develop somewhat more rapidly and a source of infection or a history of traumatism is usually present. Cerebral syphilis; the fact that syphilis of the brain may be in the form of a gumma makes the differential diagnosis in some cases extremely difficult. In the presence of positive biologic reactions or stigmata of hereditary lues a trial of syphilitic therapy may help to establish the diagnosis.

Nephritis.—Nephritis may cause increased intracranial pressure and edema. There may be at times localizing symptoms, but the presence of other symptoms and signs of nephritis will usually serve to differentiate. Chronic encephalitis; chronic encephalitis may be present in association with brain tumor. It is not likely to produce the marked increase in intracranial pressure that the tumor would cause, although cases on record show it has done so. Post-basic meningitis; Post-basic meningitis followed by internal hydrocephalus, except for the localizing signs, may produce a perfect clinical picture of brain tumor. It should be suspected in children. Acute swelling of the brain, acute serous meningitis etc. are rare conditions which cause the syndrome of increased intracranial pressure. The diagnosis is as a rule quite impossible.

The *x*-ray is of little value in the diagnosis of brain tumor, except as it shows destruction of the landmarks of the base of the skull by pressure or an increase in the convolutional markings in the skull. It is impossible to show, directly, the brain tumor by *x*-ray (Shuller).

Spinal-fluid findings may be positive in brain tumor especially if the tumor impinges upon the meninges. Usually the cell count is somewhat raised by a reaction on the part of the meninges. The cells in such a case being lymphocytic in character. Cases are on record in which tumor cells have been found in the spinal fluid. The kind of tumor cells are not necessarily exactly the same as those composing the tumor, for instance in a case of spindle-cell sarcoma of the brain, round cells were found in the spinal fluid, etc. The albumin and globulin contents of the spinal fluid may be increased and the ninhydrin reaction may show hydrolized, simpler forms of protein. Dandy has suggested that ventricular puncture followed by the injection of air into the ventricles and an *x*-ray examination subsequently is of value in the diagnosis of tumors inasmuch as it will show any distortion or blocking of the ventricular system. The results so far obtained by this method are encouraging. It is especially useful in differentiating hydrocephalus from tumor.

Prognosis.—The prognosis in tumor of the brain depends on a number of factors. The presence of teratoid tumors are not incompatible with normal brain function and continued health. Various angiomata etc., while producing symptoms, may not grow. Encapsulated tumors especially if of small size and on the surface of the brain, may be removed. The prognosis in gliomata and carcinomata is always serious. Solitary tubercles may retrogress spontaneously. Gummata may disappear under appropriate treatment.

The usual **treatment** of tumors of the brain is their operative removal. Advances in brain surgery have made it possible to remove tumors of the brain from regions formerly considered inaccessible, i.e., pituitary, pineal. Surgical skill in performing these operations, however, is not widely distributed. In many cases the attempt at removal of the tumor is attended by fatal consequences. It has been estimated that approximately 20 per cent. of all brain tumors are operable. This figure is a rough approximation and probably not more than 5 per cent. are capable of real cure. In cases of more inaccessible tumors it is often necessary to relieve the intracranial pressure by the removal of a portion of the skull bone. The site of choice for such an operation is either over the tumor or, in case of unlocalized tumors, over the right temporal sphenoidal lobe, the latter point is chosen because the temporal muscle affords some protection and also because in event of cerebral hernia, which almost invariably follows, the functional effects of the hernia will be less marked if it occurs in this region. In some cases a decompression operation for a tumor which is subcortical, has lead to the tumors growing toward the opening and its subsequent removal. Decompression should be undertaken early in cases with choked disc in order to save the eyesight. Ventricular puncture or puncture of the cisterna magna might also be useful to relieve the intracranial pressure. The effects of such procedure, however, must be regarded as generally temporary. Spiller and De Schweinitz have advocated repeated lumbar punctures for the relief of choked disc. Lumbar puncture is generally regarded as dangerous in case of brain tumor owing to the possibility of a herniation of the medulla into the foramen magnum with consequent pressure and sudden death, but where the operation is done carefully and only a small amount of fluid is withdrawn it may be carried out safely.

X-ray treatment of tumor of the brain has shown some results. In a case under my observation with a cerebellar growth, the patient regained his ability to walk. The symptoms, however, again increased. Necropsy showed the tumor to be a sarcoma. In another patient with tumor of the frontal lobe there was some improvement in the mental symptoms, but the patient subsequently died from hemorrhage into the tumor. In another case of tumor of the frontal lobe an operation showed nothing on the surface but a microscopic examination of a "brain plug," removed at the time of operation, showed a glioma about one-quarter inch beneath the cortex. This patient had six *x*-ray treatments following the operation. When examined about four months later she had no symptoms of increased intracranial pressure and no cerebral hernia. Permission to remove another brain plug for purposes of comparison was refused by the patient, but it appeared that the growth of the tumor was inhibited at least. It is possible that if the bone is removed directly over the tumor and some of the tumor removed, subsequent *x*-ray treatments or radium treatments might be valuable.

Internal medication is valuable in the gummata and the use of potassium iodid in large doses seems to inhibit the growth of other tumors as well. The use of Coley's serum in a case of sarcoma of the brain, was without effect. In another case I used ascitic fluid from a case of carcinoma injected into another patient with carcinoma of the brain. There was considerable reaction of anaplylactic type, followed by distinct improvement, however, the improvement was not permanent. Autopsy confirmed the clinical diagnosis. Symptomatic treatment, aside from decompression operation, is usually confined to the relief of head pains and the general symptoms. For the relief of headaches the coal-tar products are sometimes useful. Morphine is usually ineffective and inadvisable. It might be valuable to give injections of hypertonic salt solution in these cases with the idea of temporary relief of the head pains by reducing the intracranial pressure.

The vertigo nausea and vomiting, etc., are usually best relieved by keeping the patient entirely quiet.

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CHAPTER CLXXX

ACUTE POLIOMYELITIS

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Nomenclature.—The Bureau of the Census[1] has urged the uniform adoption of the term "acute anterior poliomyelitis." In 1909 in collecting statistics, returns were found under 24 different diagnoses. Various synonyms for the disease are: Spinal infantile paralysis, poliomyelitis anterior acuta, epidemic infantile paralysis, epidemic poliomyelitis, anterior poliomyelitis idiopathic, essential infantile paralysis, myelitis, poliomyelitis, meningo-myelo-encephalitis disseminata, Heine-Medin disease and infantile paralysis.

Definition.—Acute poliomyelitis is a general systemic infectious disease which in a certain small majority of cases tends to affect the central nervous system, especially the gray matter of the anterior horn and the ganglion cells of the motor nerves but also at times causing lesions in the posterior spinal root ganglia, resulting in paralysis in a certain proportion of the cases, either flaccid or spastic depending on whether the lower or upper motor neurons are affected.

Historical.—There is little question that this disease as we know it now, existed in sporadic form a long time before it was recognized as a separate entity. The earliest credit for describing a case is given to Underwood,[2] who in 1787 in his "Treatise on the Diseases of Children" described the condition as "the debility of the lower extremities." He describes the symptoms, attributing the condition to teething or foul bowels. "It is then a chronical complaint, and not attended with any affection of the urinary bladder or with pain, fever or any manifest disease, so that the first thing observed is a debility of the lower extremities, which gradually become more infirm and after a few weeks are unable to support the body." He says that children are usually affected, seldom under one year or over four or five years of age. Mitchell[3] describes the condition in the skeleton of an Egyptian mummy dating back to 3700 B.C.

These descriptions, however, do not bear critical analysis, as they do not differentiate between paralysis due to poliomyelitis virus as we recognize it now and that due to other lesions such as tuberculosis and birth palsies. Jörg, in 1816,[4] wrote the first description of the disease, which is a very accurate account of the acute onset. The description is as follows:[5] "The little girl was born healthy, although the mother was by no means strong. When a few weeks old the child suffered from some violent fever, of the nature of which the parents could tell me nothing, but which seemed to have been caused by a chill and to be of the nature of those typhoid fevers to which children are so subject and which make such devastating inroads on their whole economy. The illness lasted a long time, and the parents and the doctor despaired of all hope. In spite of this the child gradually recovered from the fever, although she remained for a long time wasted and weak. About the same time the mother suffered from some illness of which she very nearly died and the child consequently was left to the care of strangers When the mother had recovered sufficiently to resume the care of the child she noticed that it did not move its feet properly; it became more and more clear that the feet were becoming clubbed."

Badham[6] in 1835, began to question the cause of these paralyses. He is among the first who apparently took any interest in the condition. In his discussion he asks "What is the cause of this paralysis? Of what nature is it? What is to be done against it?" He calls upon the medical men of all nations to publish their experiences and views in this important matter. Brück,[7] in 1839, describes the case of a boy who developed contractures of all the limbs after an attack of infantile paralysis.

The earliest accurate description, however, was given by Jacob von Heine,[8] who was an orthopedic surgeon in Camstadt, at the head of the state orthopedic institute. In 1838 he first discussed the subject before the Society for the Study of Natural History at Freiburg and in 1840[9] wrote his memorable monograph in which he gives the disease the name of "Spinal Infantile Paralysis." He chose this name because the cases which he observed were either adults who had been stricken with the disease in childhood or were children lately affected. By his careful, analytical, deductive reasoning, he convinced himself that the site of the lesion was in the spinal cord. Later, as the anatomical distribution of the pathologic processes became better known, the name "Poliomyelitis anterior acuta" came into use and has persisted on account of the region which the disease principally attacks. Later, as the condition was better recognized and epidemics began to be described in various parts of the world, "epidemic infantile paralysis" and "epidemic poliomyelitis" were used.

As the pathology of the condition became better known through the hypotheses of Strumpell[10] and Pierre Marie[11] and especially through the work of Medin,[12] who proved that the conception was correct that certain forms of encephalitis were connected pathologically with the classical infantile paralysis and that the brain and the spinal cord might either one be affected, the terms so far given were shown to be more or less inadequate.

French authors applied the term "idiopathic" or "essential infantile paralysis." The term "myelitis" appeared and as the exhaustive pathologic work of Harbitz and Scheel[13] and the generally accepted classification of Wickman[14] was confirmed by all pathologists, that the disease did not limit itself to the gray matter but usually involved the meninges and frequently the white matter of the cord and brain, the term "poliomyelitis" became too narrow and the name "meningo-myelo-encephalitis disseminata" was coined. But this name is clearly too long; although it partially expresses the pathologic conditions, even it is not free from the objection that it does not include the generally infectious nature of the disease. Writers such as Wickman and Romer have adopted the name "Heine-Medin disease" and although as a general principle, there is a real objection to calling a disease by the name of a man or group of men, it is probably less objectionable than the inaccurate nomenclature of "infantile paralysis" or "poliomyelitis" which are so current in American literature dealing with this disease.

Our present knowledge of the disease is the result mainly of a few prominent workers and it is to these alone that we will make any reference though there are many who have worked on this disease and have produced more or less important changes in our conception of the disease.

A short and very good description of the on-set and acute stage of the disease is given in v. Heine's first monograph.[8] He considered that the paralysis depended on a sudden, serous exudate into the spinal cord. His description of the acute stage is worth quoting:[15] "The disease attacks children who are healthy and well developed at the age of from 6 to 36 months. Usually they have suffered from no illness previously, in some cases they have been somewhat unwell for a short time. The incidence is sudden, occurring with fever, congestion of the head, restlessness, and, in short, symptoms of general irritation. At the same time there are signs of difficulties in dentition, the children put their hands to their mouths, from which saliva pours, the gums are swollen in places and are hot, sleep is restless, broken by paroxysmal cries, and the eyelids are only half closed. Sometimes the disease begins with vomiting, diarrhea, or the appearance of acute rheumatism. Rarely one of the exanthemata seems to be at the root of the matter. Subsequently there may be more or less severe convulsions, which may recur. In two children, whom I treated later, the illness began suddenly with convulsions, collapse, frothing at the mouth and nose, and lividity. In other cases the paralysis appears after only slight general symptoms of fever which may be overlooked; the child goes to bed apparently healthy and is found paralyzed in the morning. This mild type of onset is the more common form and often leads both the doctor and the parents to take too hopeful a view of the illness. The danger to life in such cases is very slight, but there is no doubt that in the more violent cases the child is in considerable danger, although no directly fatal case has come to my knowledge. The convulsions are limited to a single attack in most cases, but rarely the initial attack is followed by several minor paroxysms. After the convulsions the child lies quiet, pale and exhausted, looking about the room as if just awakened from a sleep and it appears to the parents as if convalescence will be rapid until they find on lifting the child that it is paralyzed. The lower extremities are most frequently affected; often in conjunction with the muscles of the trunk, so that the patient not only loses the power of walking, but even of sitting up and of holding up the head. Frequently one leg is affected, but without the corresponding arm, as in the hemiplegic type of the disease; a few separate muscles may be affected in each leg. In the rarest cases one arm and shoulder being affected the limb hangs by the side, while the lower extremities remain quite untouched. The bladder and rectum are sometimes weakened for a time, but never permanently affected. With the occurrence of paralysis the first stage of the disease is finished and it passes into its second state." This description Heine mainly obtained from a history of cases. Before the days of epidemics it was rare for the doctor to have an opportunity of observing the acute stage of the disease.

For 40 years following Heine, very little work of any importance appeared. Several small epidemics were described.

Medin[16] called attention to the fact that Bergenholz had recognized an epidemic in 1881 and also that Oxholm and Cordier had each described smaller epidemics. In 1870 Charcot[17] studied the pathology in these paralyses and described a primary degeneration in the anterior horn cells. Around this very point have waged most of the early pathologic discussions.

Strümpell[18] in 1884, pointed out the resemblance between certain cases of acute encephalitis with spastic paralysis in children to cases of "infantile spinal paralysis." He, for the first time, suggested the hypothesis that an external infectious agent was responsible for both conditions. This recognition by Strümpell that there was a true cerebral form of this disease is one of the most important contributions to the study of this disease. Strümpell says: "Passing in review the other nervous diseases from this point of view, one group stands out by itself, the members of which show great similarity with one another in their course and in their symptomatology. This group consists of the acute multiple neuritis, acute poliomyelitis and acute encephalitis of children. All have a sudden onset accompanied by considerable fever. The patients show mental dullness, suffer from headache, gastric disturbances, and occasionally enlargement of the spleen, swelling of the joints, and a slight albuminuria. According to our present point of view, these are all signs of infection by a pathogenic organism." In another place Strümpell says: "The peculiar clinical appearances of spinal infantile paralysis have frequently given rise to the opinion that in this disease we have to deal with an acute infective process. As in the case of so many undoubtedly infectious diseases, we cannot prove this to be so by direct demonstration of the causative agent. At present we can rely only upon the following facts: The peculiarity of the clinical course; the sudden onset for which no obvious cause is present; the moderately high temperature; the severity of the general symptoms; finally, the nature of the changes found in the spinal cord, which point unmistakably to a truly inflammatory process." And again he writes in 1884: "Even the general course of these diseases shows great similarity. Multiple neuritis and poliomyelitis appear in similar varieties; they show the same gradation between the acute and the chronic forms. Both affect principally the motor tracts, although owing to the peripheral nerves being affected in the one and the spinal cord in the other there are many minor differences in the symptoms. It follows that the purely anatomical classification between neuritis, poliomyelitis and acute encephalitis, which is now in vogue, must be looked upon as an artificial one. It seems to us worth waile to consider whether all these diseases may not be considered to be different local manifestations of the same disease brought under one etiologic heading, and whether they may not be considered to be different local manifestations of the same disease or at least of very closely allied diseases. In this connection one might apply the analogy between croup and faucial diphtheria to the diseases, multiple neuritis and poliomyelitis. It is only of late years that the former diseases have been recognized as arising from the same cause, whereas for many years a purely artificial distinction was made between them."

In the year 1885, he writes: "Both diseases tend to attack previously healthy children in infancy. They present a stage of acute invasion in which they can scarcely be differentiated; this leaves a paralysis behind it which in both diseases affects the motor tracts, in the place of poliomyelitis the gray matter of the cord, in the case of the hemipelgic plasies undoubtedly the brain; this resultant palsy is naturally of different kinds, but the differences are due merely to the localization of the disease and not to any difference of the essential nature of it. In any case the analogy between the two diseases is remarkable in that in both cases it is the gray matter which is affected. That in the hemiplegic cases, the cortex of the brain is affected, is proved by the distribution of the paralysis, the occurrence of epileptic attacks and athetosis afterwards, and, from the anatomical aspect, by the results of microscopical examination. In these cases porencephaly is found which is not of the kind present in cases of congenital defect, but which still show evidences of its inflammatory origin, further, this porencephaly is found in the central convolutions, that is to say, in the motor regions. The appearances are precisely similar to those found in the anterior horns after an attack of poliomyelitis. There is at present no record of pathologic investigation of the acute stage of either disease.

"It seems to me to be justifiable to class acute encephalitis as a by no means rare variety of acute anterior poliomyelitis. Personally I lean towards the opinion that the two diseases are of the same essential type, or are even identical, inasmuch as both are due to the same agent, possibly of an infective nature which sometimes attacks the gray matter of the cord and at others the cerebral cortex. In order to express this relationship in the nomenclature, I should recommend that this particular form of hemiplegia should be called cerebral infantile paralysis or polio-encephalitis acuta in contradistinction to spinal infantile paralysis or poliomyelitis acuta. The diagnosis of the former is arrived at easily by means of careful investigation; at the same time the existence of hemiplegia in children due to other causes must not be lost sight of."

The first extensive epidemics which occurred were in Sweden. These were studied and reported by Medin[19] in 1887. The first epidemic of any magnitude, 43 cases, occurred in and around Stockholm that year with three deaths. In 1890 Medin[16] reported to the Tenth International Medical Congress, giving an account of his clinical studies of these cases. This account is the first good clinical picture of the disease, and it is for this reason that his name is appropriately placed with that of Heine. In 1895[19] he again reported a smaller epidemic of 21 cases. In his studies he brought out the points which Strümpell had emphasized but which had not gained much recognition, the close relationship of the spastic and flaccid types of paralysis and the involvement of the cerebral and bulbar areas as well as of the peripheral nerves. From his studies, he drew the deduction that this disease must be contagious. During the later epidemics which were carefully studied by Harbitz and Scheel, Wickman and others, we still find references to Medin as the active, keen clinician, aiding in the more intensive study of this disease.

One of the first epidemics in this country was the one reported by Colmer[20] in 1843. While on a visit to the parish of West Feliciana, Louisiana, in the fall of 1841, he had his attention called to a child about one year old, then slowly recovering from an attack of hemiplegia. "The parents (who were people of intelligence and unquestionable veracity)," he writes, "told me that 8 or 10 other cases of either hemiplegia or paraplegia had occurred during the preceding three or four months, within a few miles of their residence, all of which had either completely recovered or were decidedly improving." Colmer says that all of these cases were under two years of age and though he did not recognize the condition for what it was, ascribing it to teething, still there can be little doubt that this is the first reported epidemic in this country.

Between then and the early eighties, there were few reported epidemics. Since 1880, however, there is hardly a year without a report of an epidemic and this gradually increases until the present series of epidemics which apparently started in Norway and Sweden between 1903 and 1906. Caverly[21] and McPhail[22] reported an epidemic in Rutland, Vermont, in 1894, in which between July and September, the records of 126 cases with 18 deaths were collected. During this epidemic it was noted that certain animals, horses, dogs and chickens, were affected but no scientific work was done with this material.

J. M. Taylor[23] reported the next epidemic which occurred in Cherryfield, Maine, seven cases being noted during July and August, 1896. The apparent infectiousness of the condition was commented on as there was close family relationship between those affected, there being a brother and sister and a cousin among them. The others came in direct contact with these three. Bondurant[24] reported 15 cases in Greene County, Alabama. H. L. Taylor[25] reported a number of cases which he saw in New York at the Hospital for Ruptured and Crippled in New York in July, 1897. In the same year Sherman[26] reported eight cases which occurred during the summer of 1896 in San Francisco and Napa. In 1898 Newmark[27] reported a small epidemic which occurred in La Grande, Merced County, California. La Grande had a population of 49 people, and of this number four children were affected, all boys between the ages of 8 and 10 years. Two of them were brothers and the third their playmate, while the fourth was the son of the doctor who attended them.

Packard [28] reported two cases, a brother and sister in Royersford, Pennsylvania in 1898. In 1900, Chapin[29] reported seven cases which he saw in Poughkeepsie, New York, during July and August, 1899, and during the same summer between 30 and 40 cases were seen by other men in that locality. McKenzie[30] reported 17 cases which occurred in two small villages in the same district. Painter[31] reported in 1900 52 cases in Gloucester, Massachusetts, all in children. Woods[32] reported an epidemic of 55 cases in March and June, 1901, in San Francisco.

The seriousness of the disease and the widespread disability which results from it was, however, not clearly brought out until 1905, when the disease apparently become more serious and invaded a great many districts in Norway and Sweden. The rapid spread of the disease in these countries is shown as follows: There were 18 cases with six deaths in Norway in 1903; in 1904, there were 41 cases with six deaths in one epidemic and 20 cases with six deaths in another; in 1905, there was a total of 719 cases with 111 deaths and the following year 334 cases with 34 deaths.

The first large epidemic in the United States occurred during the summer of 1907 in New York, Massachusetts, Connecticut and other Atlantic seaboard states. Dixon and Karsner[33] reported the occurrence of 200 cases in northwest Pennsylvania. The disease began to show itself in May, 1907, and the cases increased rapidly during June and July, the disease reaching its heighth in July and August. During this epidemic there were at least 2500 cases of the disease in New York and vicinity. A careful study of the New York epidemic of 1907 was made by the collective Investigation Committee,[34] and the same epidemic was investigated by Lovett[35] and Emerson.[36] Since 1907 the disease has been endemic in this country. Three other epidemics were reported during this year, that of Terriberry[37] in Ridgway, Pennsylvania, 50 cases; of Griffin[38] in Oceana County, Michigan, 20 cases; and the third reported by Free[39] in the Alleghany Mountains. During 1908 Jacolyn Manning[40] reported an epidemic of 352 cases in Wisconsin and Hamilton[41] reported three epidemics which occurred in Minnesota. Lovett,[42] in a special report on infantile paralysis to the Massachusetts State Board of Health, says that in five years previous to 1909, there were about 8054 cases reported in the world; 5514 of these cases occurred in the United States, chiefly in the northern states.

During 1909, Massachusetts, Nebraska and Minnesota apparently suffered the heaviest toll; Shidler[43] and McClanahan[44] reported epidemics in Nebraska. An epidemic occurred in the State of Washington in 1910, during which time there were 397 cases.[45] In Massachusetts the disease was nearly as prevalent during 1910 as in the previous years. O'Reilly[46] reported the occurrence of 309 cases of infantile paralysis in Missouri in 1910. During this same period (1910) there were extensive epidemics in Iowa, Nebraska and Kansas; and a large epidemic (125 cases) in California which was reported by Fleischner.[47] A small epidemic in Portland, Oregon, was reported during the winter of 1909 to 1910.[48]

During 1911, poliomyelitis became epidemic in Ohio and increased rapidly through the next few years. During 1913, it made its appearance in central Alaska on the Yukon River where Pierson[49] reported an epidemic. Sporadic cases had preceded this epidemic but this was the first large epidemic. Pierson also refers to a like condition among dogs which was considered to be distemper.

According to the United States Census Bureau over 5000 deaths from poliomyelitis occurred between 1910 and 1914 in the registration area. Emerson[50] says: "A conservative estimate would place the probable number of cases during these five years in this area as 30,000."

The largest epidemic this country has ever known occurred in 1916, over 27,000 cases being reported in the registration area from June to November in 1916 with 6000 deaths for this single epidemic. The main site of this epidemic was in New York state where 13,164 cases occurred during this year, most of these cases being in New York City, only 4165 cases having occurred outside the city. Since that time various small epidemics have occurred in different sections of the country but no very extensive epidemic has been reported up to 1921 since 1916 in the United States.

Epidemiology.—The mode of transmission of the epidemic type of acute poliomyelitis has been extensively studied since Wickman[14] first drew attention to the contagious character of the disease in 1905. Wickman first called attention to the abortive and meningitic forms of this disease and pointed out the probability of carriers. Up to then, the paralytic type was the only type of the disease recognized and cases without paralysis were not regarded as belonging to this group. When we consider that in epidemics, paralysis may be entirely absent in quite a large percentage of those affected, it is clear that the mode of transmission of the infection must be considered in a different light than it has been heretofore. (From 50 to 80 per cent. of cases are supposed not to show paralysis.) It follows from its epidemic character that poliomyelitis must be an acute infectious disease and we know of no disease that flourishes in epidemics that is not due to a transmissible cause. That not everyone is affected by paralysis that has the infection is shown by many families or households studied, where the occurrence of dual or multiple infections in one family or household has not been so uncommon as it was believed to be. We now recognize the abortive and meningitic forms of the acute infection and the unaffected innocent carrier and realize that the results of the infection may show themselves in either a spastic or flaccid paralysis, depending on whether the upper motor neuron or the lower motor neuron is chiefly affected.

The presence of abortive cases is well illustrated by the following history taken from the monograph of the Rockfeller Institute "A Clinical Study of Acute Poliomyelitis" by Peabody, Draper and Dochez.[51]

Family 3.—"Hannah T., age four years. On October 1, she was well in the morning but vomited in the afternoon and was feverish. The next day she was paralyzed. There are four other children in the family, one older and three younger than Hannah. One week before her illness the older brother had had tonsillitis, and a day or two after his attack began, the three younger children had what the mother called a "drowsy sickness," in one case with fever. A small brother, one of the three, was brought to the hospital with typical symptoms of the acute stage of the disease and significant changes in his spinal fluid. There were no definite paralyses, but a distinct shakiness in walking was noticed."

These authors say "These family infections speak for themselves in the matter of direct contagion. But there are many factors that make one hesitate in accepting the contagious nature of the disease, at least, we must have more light on the way in which the disease is carried from one individual to another before we can come to any definite conclusion. Its contagiousness, at the most, is considerably less than many of the contagious diseases which we have with us in endemic form. The last instance quoted, though lacking definite proof, is highly suggestive of the importance of abortive cases." One further example, however, quoted from the same monograph, where the evidence of the neutralization test made probable the diagnosis of acute poliomyelitis, in a case that had no muscular weakness, is very interesting.

"Helen K., age three years and ten months. On August 16, she had headache and was feverish; on the eighteenth, paralysis of the legs began and ascended rapidly to the arms and terminated fatally by respiratory involvement on August 21. Her constant playmate, Helen B., age six years, had headache on September 1, that is about 10 days after Helen K. was ill, and the following day was brought to the hospital. Examination of the spinal fluid showed changes similar to those seen in poliomyelitis. She never developed paralysis and after 15 days went home apparently well. Her blood serum protected a monkey against 0.1 c.c. of the virus.

"This child had no definite symptoms beyond some stiffness of the neck, headache, apathy and anorexia. Except for her association with the fatal case, her mother would have paid no attention to what might have passed as a slight summer ailment, and a diagnosis would never have been made. The child would have gone to school in a few days and perhaps have been the source of infection for other individuals."

The variability of infection to contagious disease is well known to us. We know that scarlet fever and typhoid are escaped by more people who come in contact with these infections than are escaped from small-pox and measles. Moreover, the contagiousness and infectivity of the various communicable diseases have very definite seasonal or yearly variations.

Most epidemics of acute poliomyelitis have occurred during the summer months, from June to September or October. In the New York epidemic the heighth of the epidemic was reached during the middle of August, in New York City a week earlier than in the state as a whole (August 7 to 13 as against August 14 to 20 throughout the state). However, winter epidemics have been described by Leake,[52] who reports 39 winter outbreaks, though as he points out the epidemic in Elkins, West Virginia, during November, 1916, undoubtedly was due to the New York epidemic of the summer of 1916. Most of the winter out breaks have been comparatively small. The only one in which there were over 100 cases was that in Vienna and lower Austria from November, 1908, to February, 1909, in which there were 125 cases. The variations are also quite marked as is shown by the number of cases in New York since 1910.

TOTAL NUMBER OF CASES OF POLIOMYELITIS[53] IN NEW YORK STATE SINCE 1910

Year	Cases
1910	112
1911	139
1912	1,108
1913	491
1914	224
1915	257
1916	13,164 (4155 total cases excluding New York City)

Frost[54] considers the infectivity of poliomyelitis to be only one-fifteenth of that of scarlet fever though as has been pointed out a susceptibility to one of the less contagious diseases indicates that the child is more apt to contract other contagious and infectious diseases. In the New York epidemic of 1916 there were 8635 families involved, 96 per cent. of these had only one case; the total number of children in these families were 24,883. There were 3.6 per cent. of the families which had two cases and 0.3 per cent. had three cases and 0.06+ per cent. had four cases, so that multiple cases only occurred in 4 per cent. of the families involved.[55]

Regarding the degree of contagiousness of poliomyelitis, an interesting comparative table is given in the Monograph of the New York City Department of Health[55] dealing with this question, comparing it with the incidence of diphtheria, scarlet fever and measles as occurring on the East Side of Manhattan, which has a total population of 430,000.

COMPARISON OF INCIDENCE OF CONTAGIOUS DISEASE, CORLEARS DISTRICT

	January	February	March	April	May
Diphtheria	132	103	122	123	139
Scarlet fever	41	36	35	59	66
Measles	25	58	186	287	288
	June	July	August	September	October
Poliomyelitis	13	189	154	20	4

Leegaard,[55] in 1899, first called attention to the fact that infections apparently spread along lines of travel. Since that time, this has been the proven theory in epidemiology. All evidence seems to point to the fact that epidemics have traveled along great arteries of commerce and travel, along waterways and railroads. There is a tendency, undoubtedly, to grouping of the disease. Wherever case studies have been carred out, intimate intercourse of households and meeting places have been the main ways of spreading the disease. Wickman called attention to this. His explanation apparently still holds that the disease spread like other contagious diseases and that the abortive case and the healthy carrier are the most important factors in the transmission. Wickman considered, and it is still generally held, that indirect transmission by non-living objects is very uncommon. However, some facts, especially during this last epidemic in New York (1916), seem to indicate anew the possibility of the contagion being carried by other than human agents.

Brues,[57] after investigating the possibility of insects as carriers of infections, states that "Many facts of distribution and general prevalence suggest that the disease is at least to some extent dependent upon some population other than the human one for its spread. Insects migrating by themselves, or on the body of some animal host like a rat, might easily account for the facts dealt with in this section of the report." He thinks that the most possible insects may be fleas which are harbored on rats and cats or it may be due to flies, especially the stable fly.

Lately Saunders[58] has called attention to the work which he and his associates have been carrying on with the Lucilia Caesar fly, which he terms the "paralysis fly." This fly is a carrion fly which deposits its eggs on decaying animal and vegetable matter, especially upon excrement, and he notes that the lone "Lucilia Caesar delights to deposit its ova upon man's food, likewise into any orifice of the human body—the ears, the nose, the lips, the anus, and more often upon the milk ducts of mammalians. Hence the frequent occurrence of paralysis in sucking animals." His experimental work also goes to prove that this fly may be a carrier of the virus. However, in all of these cases, the possibility of abortive cases and healthy carriers is hard to rule out.

The experience of the New York City Department of Health in isolating children of institutions is most illuminating.[59] They isolated 93 institutions for the care of children, such as asylums and children's homes, which had a total census of 21,746 children, and 76 institutions for the temporary care of children which had a total census of 6365 children. These institutions were under strict control and supervision of the Department of Health. From a careful study of these institutions they conclude that "Isolation of groups of children from contact with other children or adults, even when carried out in the midst of areas where the disease is prevalent, suffices to protect such children almost absolutely from infection in spite of the use of identical water and food supplies and exposure to the same atmospheric conditions and winged insects." The few cases which occurred in these institutions showed that the infection existed previous to their admission, no cases developing among children who had been in the institutions for longer periods than the accepted incubation period of two weeks and where visitors were excluded. This is very strong evidence for the isolation of children during an epidemic.

Two additional examples of the efficacy of isolation are as follows:[60] "There were between 80 and 90 children on Governor's Island, the United States Government Military post, throughout the epidemic. They were living under as nearly ideal sanitary conditions as may well be obtained. Absolute exclusion of all children visitors to the island was maintained from July 4. The children of the island were not allowed to leave until the middle of September. No case developed."

The second example: "There were 350 children, under 16 years of age, on Barren Island in Jamaica Bay, Borough of Brooklyn. To this island all the city garbage and offal is taken for reduction in the large rendering plants. Flies and mosquitoes are abundant. Rats are numerous. There is no public water supply and there are many shallow surface water wells. There is no sewage system. There are few, if any, cellars. There is no garbage collection. There are no public highways. The population of about 1300 people represents the lower grade of unskilled labor, Poles, Italians and Negroes predominating. The standard of living is low. No cases of poliomyelitis developed on Barren Island."

Römer[61] cites a very interesting case, showing how the disease is spread:

"A particularly instructive example was provided in the case of the small epidemic in the little town of Frankenau. The midwife of the town had visitors from Westphalia from September 11 to 13. On the twentieth, one of her children was stricken with paralysis and two others suffered from gastro-intestinal symptoms. Sometime later a neighbor's child, who had played with the children of the midwife who had remained unaffected, became ill, so also a child of the school master (the healthy children of the midwife attended the school); finally, the child of a workman became ill, who had been in the same schoolroom with the midwife's children. The track of the epidemic was therefore as follows: The virus was brought by healthy carriers from Westphalia; in the village it was disseminated by the healthy children of the midwife."

The importance of carriers becomes more evident as we study the course of the disease and at present offers the best explanation of the spread of the disease. There are some factors, however, which are not explained by this hypothesis. The carrier is probably a larger factor in distributing the infection than are the cases which are paralyzed or confined to bed during the active period. We know that very much the same is true of other contagious diseases, as diphtheria and scarlet fever, for although these, like paralysis, are not highly contagious, they are undoubtedly spread by healthy carriers. As far as our present knowledge goes, we must accept the probability of the human agent being the most important in the transmission of the disease.

Paralytic diseases among animals, especially domestic, are not infrequent and are especially frequent among barnyard fowls and are often noticed among household pets, dogs and birds, and among horses, and occasionally it is met with in young rabbits. All forms of these paralyses have been studied by various workers from the experimental standpoint but so far they have not been able to reproduce the typical paralysis in monkeys by inoculation experiments nor are such paralyses transmissible by inoculation from chicken to chicken or chicken to monkey. The pathologic lesions characteristic of poliomyelitis have not been found in these cases of spontaneous paralysis among animals. The pathologic lesions found in such cases are definite lesions of the peripheral nerves. These peripheral neuritises are more often due to unusual and improper forms of food, as is shown by the work of Theobald Smith and Flexner and Clark. The fact that it has not been possible to transmit the paralysis from one animal to another, however, does not prove the possibility that the virus may not exist among domestic animals because they do not respond to this particular method of transference or that the lesions are so different that they are not recognized as poliomyelitis in man. This is made especially clear by the confusion which has arisen from the experimental work on rabbits. It has been demonstrated and now accepted, that young rabbits sometimes, but by no means always, respond to the inoculation of the virus of poliomyelitis. Older rabbits are not susceptible to inoculation. This experimental paralysis in rabbits, however, often only shows itself by general weakness and the pathologic picture and findings are often different from those in the monkey or in human cases. Different types of experimental paralysis in the rabbit should be differentiated from the type of peripheral neuritis which occurs spontaneously in very young rabbits. This is probably true of some other type of infection than poliomyelitis.

Other means of transmission than from individual to individual have been demonstrated but most of these act in a purely mechanical way, such as the stable fly, which has been shown to be capable of transferring the virus, but this is probably true only under experimental conditions and has never been shown to do so under ordinary conditions. In fact even under experimental conditions it is difficult to transmit the virus by this means. In the same way, dust[62] from a sick room has been shown capable of reproducing the disease. This transference, however, is strictly passive as the virus may continue in a living state, actively infectious for 48 hours or longer. For the time being, at least, we must consider the human carrier, active or passive, as the main source of infection. However, until this part of the epidemiology has been more definitely settled, one must remember the early theory of contagion in such infectious diseases as scarlet fever and malaria. It is interesting to quote in this connection from Allbutt's "System of Medicine"[63] (1911).

"Of the epidemic character of yellow fever, more is known. When the infection has been introduced into a locality it usually appears in single or isolated groups of cases for the first two or three weeks or longer, and these are restricted to a particular district or districts. In this, as in other respects, it reminds us of the plague. After the disease has once escaped these limts it spreads with a rapidity proportionate to the density of the susceptible population, to the freedom of intercourse between the healthy and the sick, and to the degree of temperature; and its intensity is specially favored by overcrowding, want of ventilation and, according to some, by an impure, undrained soil, and insanitary conditions.

"When the contagion of yellow fever has been introduced into a non-endemic area the disease is spread by intercourse with the sick or with contaminated persons, themselves healthy, by means of articles charged with the infection; by residence in or visits to infected houses; and it may be further propagated by aerial diffusion from house to house, if in close contiguity. The evidence of its communicability by means of water or food, or by the agency of mosquitoes, is inconclusive.

"It is right to say that these views are by no means universally accepted. As Hirsch points out: 'The most intimate kinds of contact—such as the healthy and sick sleeping in one bed, the attendance of physicians and nurses upon the sick, the use of unclean linen, clothes or beds of yellow fever patients, post-mortem examinations of their bodies, and the like—have in no wise contributed to the spread of the disease.'"

Pathology.—The older pathology dealt entirely with the late changes in the cord but with the work of Harbitz and Scheel, Wickman and Strauss, a new pathology came into being, which emphasizes not only the earliest morphologic changes in the cord but also the general nature of the infection. The changes in the cord explained the development of the later chronic forms already well known.

The meninges show the earliest changes in a very marked hyperemia and in the collection of numerous small mononuclear cells, probably lymphocytes, collected in the perivascular lymph space of the blood-vessels of the leptomeninges. These lymphatic spaces surround the vessels and are anatomical processes of the arachnoid space and the lymph in them is in communication with the cerebrospinal fluid. This probably explains why we often get changes in the spinal fluid before there is any affection of the cord. These first changes are not associated with fibrin formation or with any exudate on the surfaces of the meninges but it is more to be regarded as an acute interstitial meningitis. It is usually more marked on the anterior surface of the spinal cord and especially in the anterior fissure from which the larger vessels enter the cord. Small collections of lymphocytes are found along other meningeal vessels situated laterally and posteriorly. The blood supply of the cord is derived from the vessels of the meninges and other veins of the pathologic processes. This perivascular infiltration follows along the vessels after they enter the cord from the meninges.

The central nerve lesions in the acute stage show the meninges edematous and injected with only a moderate increase in the spinal fluid. The brain as well as the cord on section have a moist translucent edematous appearance and the gray matter of the cord is swollen, so that it projects above the level of the white matter. It is also darker than normal in color and has a rather typical grayish-pink appearance. Very often minute hemorrhages can be distinguished, more often in the gray matter but also in the white.

In both the human and experimental sections, the earliest changes in the cord are a hyperemia and the collection of small round cells in the lymph space surrounding the vessels. This cellular exudate often forms a sheath completely surrounding the vessels for a long stretch in the form of a thick collar which undoubtedly presses on the lumen and thus exerts a mechanical obstruction to the circulation.

Later, the intima of the vessels may also show hemorrhage and edema which may be either due to toxic or mechanical factors. Whichever it may be, the three factors, edema, hemorrhage and cellular exudate, are dependent on vascular changes and may be regarded as the primary reaction of the nervous system to the virus of poliomyelitis. The changes which we find in the nerve cells themselves may be due either to the vascular disturbances or the direct action of the virus. Whichever it may be, the fact remains that the vascular system is the most important determining factor in the nerve lesions. The blood supply to the cervical and lumbar enlargements of the cord and to the anterior horns of the gray matter in general is more abundant than to the posterior horns or to the white matter. This explains why these regions are most affected as their blood supply is most abundant. The irregularity with which the vessels supply the cord, the fact that the vessels are given off at different levels and supply various levels of the cord, explains why the lesions are asymmetrical and that few cases correspond absolutely in the total extent of their involvement though the general picture of the involvement is much the same when these types of paralyses are analyzed together. The clinical and anatomical pictures are both readily explained by the presence of the circulatory disturbances, the edema, hemorrhage and exudate. It has been as yet impossible to determine whether the virus may exert some direct toxic action on the nerve cells. The mechanical hypotheses did assume that the nerve cells were entirely affected by the damage produced through the three fundamental changes, hemorrhage, edema and exudate and also through the anemia which follows the constriction of the blood-vessel so that the cellular degeneration which is so typical of the later stages may be accounted for entirely by the pressure and resultant anemia. Where the hemorrhage and exudate are absorbed rapidly, cells recover their function. If, on the other hand, the anemia and pressure are prolonged or are excessive the nerve cells go on to complete necrosis and all stages of degeneration are seen in these nerve cells.

During the past few years, increased attention has been paid to the very definite and constant changes which are found in other viscera. The fact that they were hitherto overlooked or disregarded accounts for the fact that up until the last few years, anterior poliomyelitis was not considered as essentially a general infection. However, both human and animal autopsies in early cases show constant changes, more or less extensive in the parenchymatous organs and the lymph tissue throughout appears to react to the virus. Peyer's patches and the mesenteric lymph glands are usually enlarged. At times all the glandular system seems to be involved, especially the cervical, axillary, bronchial, the pharyngeal, and adenoid tissue, the tonsils and the substernal glands are found enlarged. The spleen is frequently somewhat enlarged and on section the Malpighian corpuscles are raised, pale and obviously translucent. The thymus at times shows changes identical with the lymphoid tissue. The histologic picture is almost that of typhoid fever. The lymphoid nodules show a zone of lymphocytes surrounding a more or less circumscribed pale center which is made up of large endothelial cells with oval nuclei. The nuclei are often pale and the protoplasm can hardly be distinguished. The cells are packed together and often form compact masses. Cellular inclusions consisting of necrotic cells show phagocytosis. Polymorphonuclear leukocytes appear later in the areas of extensive necrosis. Phagocytic endothelial cells are found in large number in the lymph sinuses, where they seem to proliferate. Central necrosis in the lymphatic nodules is a prominent feature as well as proliferation of the lymph sinuses.

Of the parenchymatous organs, the liver shows the most striking changes and resembles the changes in typhoid fever, where circumscribed areas consisting of a few cells, up to areas which include one-eighth of a whole lobule, in which degeneration of the liver cells, infiltration of lymph cells and polymorphonuclear lymphocytes has taken place.

The lesions are closely associated with the blood-vessels chiefly with those of the portal vein but also with the central and sub-lobular veins. Instead of the usual central necrosis, at times there seems to be peripheral spread of the lesions. The lesions vary in size and age. These lesions, degenerative and proliferative, are probably not permanent, as the liver has such a high power of regeneration. These important findings, which are constant in early human cases and in the experimentally produced disease in monkeys, make it emphatic that we recognize the disease as one which is generalized, affecting the parenchymatous organs and the lymphatic tissue, but having a specially selective preference for the nervous system. The work of Flexner and his co-workers has clearly demonstrated the importance of the general invasion of the virus and has proved its presence by animal inoculation of glandular tissue and has shown that the virus apparently comes to rest for a time in organs outside the central nervous system.

It is difficult to prove whether the visceral lesions are caused directly by the virus or whether they are secondary results of toxic substances produced during the proliferation of the virus in the central nervous system. The evidence, however, is strongly in favor of the fact that the changes are due to the virus, as the virus is found with almost as much constancy in the tonsils and nasal mucosa as in the nervous system, and as Flexner and many other workers have pointed out, organs of the upper respiratory tract play a most important rôle in the conveyance of the virus into and away from the central nervous system. The virus is regularly present in the tonsils and nasal and pharyngeal mucosa, in fatal cases in children and in infected monkeys. Monkeys have been infected by rubbing the virus on these tissues. The virus has been found in non-fatal cases in tonsil tissue as well as pharyngeal nasal secretions. It has also been found in individuals who have been associated with active cases, the parents and attendants, so that it must be emphasized very strongly that the upper respiratory mucous membranes provide for the ingress and egress of the virus of the disease, through which infection is produced and the renewal of the cause of the disease maintained.

The later picture of the process in the central nervous system shows marked degeneration and necrosis of the nerve cells; the protoplasm shows nothing but granular detritis, polymorphonuclear neurophages entering these nerve cells and completely disposing of them. In severe lesions, nothing but hyperemia, perivascular infiltration with hemorrhage, edema and diffuse cellular infiltration throughout the gray and white matter are seen, nerve cells being completely absorbed. These findings are more constant in the anterior horn of the gray matter. These same destructive changes are found to a less degree in the brain, medulla and pons. Cellular infiltration may be found in the vessels of the cerebrum and cerebellum, but usually not extensively enough to produce motor symptoms, though a marked cellular exudate and hemorrhage have not infrequently been found in the medulla and pons. Clinical symptoms and the pathologic findings often do not agree when the lesions are in the pons.

In cases which show bulbar paralysis, the posterior root ganglia show very characteristic lesions, infiltration of the small round cells in the lymphatic spaces surrounding the vessels entering the ganglia from the meninges, followed by diffuse exudate of cells, degeneration and necrosis of the nerve cells and finally, the entrance of polymorphonuclear leukocytes into the necrotic cells. These lesions in the sensory ganglia may in part account for the pain which is such a constant feature of the acute stage of the disease and another element in the production of pain is the cellular infiltration which is found along the nerve roots.

The next stage was to demonstrate the infectious agent and this Landsteiner[64] and Popper did in 1909 by producing the disease in monkeys. The following is an abstract of Landsteiner's results taken from the Zeitschrift für Immunologie, April 5, 1910:

"The material used was the spinal cord of a child of nine years, who died on the fifth day of an attack of typical Heine-Medin disease. An emulsion was made and this was injected into the peritoneum of two monkeys. The first, a young Cynocephalus hamadryas, became seriously ill on the sixth day and died on the eighth. At the autopsy the internal organs were found to be normal with the exception of the spinal cord which was seriously affected. The conditions found were: Infiltration of the pia mater of the cord with cells, perivascular infiltration in the gray matter, diffuse infiltration in the gray matter which was disorganized by edema; the anterior horns were much more affected than the posterior ones. Similar inflammatory infiltrations were found in the medulla, pons, midbrain and cortex. The cells of the infiltration were mostly lymphocytes. Where inflammation was present the ganglion cells were much altered, and chiefly in the anterior horns (outline blurred, granular disintegration, vacuolization, nuclei staining poorly, invasion by round cells).

"In the second monkey, a young Macacus rhesus, between the twelfth and seventeenth day paralysis appeared, which became complete in both lower limbs. The changes found were similar but less marked to those obtaining in the first monkey. The animal was killed on the nineteenth day. Two more monkeys were injected with emulsion of the spinal cord of this monkey, but the experiment was without result."

During the same year Flexner and Lewis published their successful transmission experiments,[65] of which the following is an abstract:

"Our efforts to transmit the disease to lower animals were first made in 1907, at which time cerebrospinal fluid obtained by lumbar puncture was introduced into the spinal canal and peritoneal cavity in monkeys and other animals. We were limited to this fluid, as we did not secure material from a fatal case. The results were negative. Since September of this year we have secured suitable material from two cases of poliomyelitis in human beings. . . . The first patient died on the fifth or sixth day after the appearance of the paralysis, which affected the lower extremities. The lumbar cord was obtained in a sterile condition, 26 hours after death, and a portion was inoculated into monkeys about 12 hours later. The entire spinal cord was obtained from the second case 12 hours after death, and inoculation into monkeys was made four hours later. In the second case death occurred on the fourth day; the lesions were diffuse throughout the cord. Paralysis had been very extensive. The gross and microscopic lesions were characteristic in both cases.

"In order to favor the transmission of the disease to monkeys, the brain was chosen as the site of the inoculation, which was made under ether anesthesia through a small trephine opening. After the operation, the animals were at once lively and normal. The injected material consisted at first of emulsions in salt solution of the spinal cord from the children and later of emulsions of the spinal cord of monkeys developing the paralysis. . . . .

"It should incidentally be mentioned that not only is the spinal cord active, but the cortex of the brain also. A delayed or unsuccessful inoculation may be converted into a successful infection by re-inoculation with an active virus."

As so often happens when the attention of many scientific workers is focused on one problem, the successful results appeared almost simultaneously. A large number of reports appeared in November, 1909. Flexner and Lewis in the Rockefeller Institute reported their successful inoculations on November 13,[65] Leiner and von Weisner[66] on November 18, Landsteiner and Levaditi[67] on November 27 and during the same month Romer[68] reported his experiments before the Society of Marburg, and in June of the following year Strauss and Huntoon[69] reported their work. These experimental studies placed the disease on a very definite scientific basis by having clearly demonstrated that Heine-Medin disease is produced by a filterable virus that shows a selective preference for the central nervous system and is not caused by bacteria.

The method of injecting the virus intracerebrally was of great scientific importance inasmuch as it gave the practical method for being able to reproduce the disease from one monkey to another. This proved that the virus was living and not merely a toxin.

Nature of the Virus.—The virus belongs to the variety which is capable of passing through porcelain filters which keep back ordinary bacteria. The virus in this filter has been sought by many methods. Romer, Joseph and Siebert[70] found by means of ultra-microscopes, small, oval slightly illuminated bodies which were motionless or showed only Brownian movements. They did not find them in normal brains. Flexner and Levaditii and Leiner and von Weisner described small round bodies. Wickman[14] drew the analogy between poliomyelitis and rabies and tried to find an increase in the Negri bodies of rabies, but failed to do so. Joest,[71] however, found intracellular bodies in the ganglion cells in cases of Borna's disease, which is an epidemic disease occurring among horses, producing a disseminated, infiltrative myelo-encephalitis, which is very similar to Heine-Medin disease in its histologic appearance, although it occurs in the olfactory lobe of horses.

However, no one has been able to find such intracellular bodies in poliomyelitis material. Romer[5] states that Leiner and von Weisner, Landsteiner and Levaditi, were all unable to find any intracellular bodies. Bonhoff[72] found bodies included in the nuclei of the glia cells. This finding of Bonhoff has not been followed up, so that it cannot be stated what these bodies were.

The effect of passing the virus through animals apparently for a time increases the virulency which after a number of passages comes to its maximum virulency at which it remains constant for a considerable length of time. Flexner and Amoss found this to be from two to four years. Then it rapidly diminishes in virulency to the point below which it started when first inoculated into monkeys.

The virus appears spontaneously in man but as far as our observations have been carried, it is not spontaneous in animals and can only be reproduced by direct inoculations in monkeys and rabbits. The experimental attack in monkeys and rabbits, especially in monkeys, corresponds very fairly with those found in man. Both the symptoms and the course of the disease, as well as the pathologic lesions, are less typically reproduced in rabbits, that is, compared with the lesions in man. Nevertheless, they are perfectly definite and typical for the rabbit.

The virus is weakened by passing through various filters, such as the Berkefeld, Chamberlain and Reichel filters. In experimenting with filtered and unfiltered virus, it has been shown that the incubation period is considerably longer for the filtered than for the unfiltered virus. That the filtrate contains the virus can be shown by reproducing the paralysis in a second and third generation of monkeys. The virus remains active in the cord for a long period. Material proof of this is rather scarce but Leiner and von Weisner found potent virus in the cord of an animal which had been ill 24 days; Romer, one that had been sick 23 days. On the other hand, virus has been found to be absent from the cord of a monkey that had suffered only six days from paralysis. Landsteiner and Levaditi quote cases in which virus became inactive after 39 to 45 days, so that no rule as to the viability of the virus in the cord is available. Virus has not been recovered from the cords of recovered human cases, which have later died from other causes and been examined for the presence of the virus.

The virus remains for a long period, however, in the mucous membrane of the pharynx, as has been shown by a number of American investigators, Flexner and Osgood and Lucas, for at least six months in monkeys and probably for as long as two years in human cases.

The virus of poliomyelitis has been shown to be a very resistant one. It withstands 50 per cent. glycerination for a long time. Cords in glycerine with 0.5 per cent. of carbolic acid can be kept without injury in this way for years. Freezing is resisted. Cords and other material have stood freezing from -2 to -4°C. for at least 40 days. Virus, however, is less resistant to heat and it is destroyed by a temperature between 40 and 45°C. for one-half hour. It apparently has little resistance to some of the milder antiseptic solutions as it is regularly destroyed by hydrogen peroxide in a 2 per cent. solution and by menthol oil and corrosive sublimate, but $1/1000$ thymol at room temperature apparently does not affect the virus in one hour. The virus, however, is destroyed by a 2 per cent. solution of potassium permanganate in one hour. With calcium solutions, 0.5 to 1 per cent. solution acts on the filtered virus more easily than on the unfiltered and destroys it in from four to five days. Drying apparently does not kill the virus. This is of considerable importance in the handling of cases.

The evidence that the upper respiratory tract is the site both for the ingress and egress of the virus has been clearly demonstrated by experimental work. The lymphatics of the nasal mucosa are in almost direct connection with the subarachnoid space. This has been brought out by Flexner in his work with diplococcus intracellularis (epidemic cerebral spinal meningitis), and by Weed in his experimental work on the flow of the spinal fluid. When the monkey is injected by an intraspinal injection of cultures they are subsequently found both in leukocytes in the tissues and free in the secretion of the naso-pharynx. Weed found that when he injected Prussian blue intraspinally that he was able to recover it very rapidly from the nasal secretion and that also it was to be found in the lymphatics of the cervical chain. The virus of poliomyelitis has been found in the nasal mucosa of infected monkeys months after an acute attack, showing that the naso-pharynx is a path of discharge for the infectious agents. A successful inoculation of the naso-pharynx of monkeys with poliomyelitis virus has shown further that the same path may serve to admit the infecting agent. Besides the naso-pharyngeal mucosa, Landsteiner and Levaditi,[67] Flexner and Clark,[73] have shown that tonsils from fatal cases also contain the virus. Similar experimental work by Osgood and Lucas[74] has proved this, as has the work of Kling, Pettersson and Wernstedt.[75]

These authors have shown that the virus can remain in the naso-pharyngeal tissue for at least six months. It is significant that the virus has been found in the washings from the naso-pharynx of parents, attendants and friends, and Kling, Pettersson and Wernstedt have found the virus present in pharyngeal swabs and washings as late as 204 days (seven months) after the infection. The longest case, the probable duration of a carrier was two years and three months (Lucas and Osgood[74]): R. H. (patient of H. G. Rockwell of Amherst, Mass.), aged five years, had his attack of paralysis in February, 1910. He was first seen by one of us (R. B. O.) Feb. 19, 1912. At that time he showed a residual paralysis of the dorsal flexors of both feet and of the peroneal group on the right. He improved until September, 1912, when after an attack of apparent bronchitis and coryza, he became suddenly weak in the right arm, with increased weakness of the affected legs as well. This was associated with fever, and the mother considered it to be a second attack of paralysis. The boy gradually recovered. Within two weeks after this apparent exacerbation his little sister had high fever, and one of her arms became completely paralyzed, with weakness of the legs. When the boy was seen on Nov. 8, 1912, six weeks after his recovery from the second attack, he was almost back in muscular strength to where he had been before, without any remaining paralysis of his arm. He still had a certain excess of nasal secretion. Through the efforts of his parents we were able to obtain 10 c.c. of clear mucoid nasal secretion on November 11. With this material passed through a Berkefeld filter, we inoculated two monkeys on November 15 with negative results. Again on Jan. 3, 1913, we received some nasal secretion and inoculated two monkeys with negative results.

We repeated this again, on January 22, inoculating 10 c.c. of the filtrate intracerebrally into monkey 157. This monkey became paralyzed February 2 and died February 5. He developed complete paralysis of all four extremities. The pathologic picture of his cord (Dr. H. C. Low) was not typical but very suggestive. On January 31 we again repeated the injection of filtrate from a fresh specimen of nasal secretion, inoculating two monkeys, Number 159 and Number 160. Monkey 159 received 5 c.c. of the filtrate intracerebrally and 10 c.c. intraperitoneally. He became paralyzed February 5 and died completely paralyzed on February 8.

Cord and medulla pathologically (Dr. H. C. Low) were typical of anterior poliomyelitis; and on April 9 we injected two monkeys, one with the cord emulsion of monkey 157, and the second with the cord of monkey 159. This last monkey, 161, received 8 c.c. of a cord emulsion from monkey 159. He became paralyzed April 27, with typical complete paralysis. The pathologic findings were typical of anterior poliomyelitis.

This case seems of special interest; first, as demonstrating the long period during which the virus undoubtedly was harbored in the naso-pharynx; second, a human infection (his sister) occurring two years after his primary attack during what seemed like an exacerbation of his former attack; and third, the recovery of the virus from his nasal secretions four months after his second attack and two years and three months after his first attack; fourth, the fact that the successful inoculations were done with filtrates from straight nasal secretions, not from washings.

The virus is unknown apart from infected human beings and monkeys although it possesses a high degree of resistance both to cold and ordinary degrees of heat for long periods. It withstands drying very well if enclosed in albuminous matter, which probably accounts for its prolonged vitality in the naso-pharyngeal tissue. It can stand both moist and dry conditions and can therefore be carried into the upper respiratory tract as a spray produced by coughing, sneezing and loud speaking, and as dust. It is evident from all the experimental work that other agents than man play a minor part in the distribution and that epidemic poliomyelitis must be considered a human borne contagious infection with the portal of entry in the upper respiratory tract, especially in the nasal pharyngeal mucous membrane. Of course, passively it can be carried by becoming attached to bedding, clothes, pets, insects, and dust.[76]

Increasingly through all these epidemics it has become more and more evident that the abortive case is of common occurrence, certainly very much more common where there is an epidemic. These cases of poliomyelitis are very difficult to recognize clinically and a specific pathologic test would be of assistance in the diagnosis of such atypical infections. But so far, the studies of Wollstein,[77] Gay and Lucas,[78] as well as others, have shown no characteristic reaction demonstrable during the acute stage of the disease. Levaditi and Landsteiner,[79] Römer and Joseph,[68] and Flexner and Lewis[80] have shown that the serum of recovered monkeys is able to protect normal animals against fatal doses of the virus this is called the *Neutralization test.* Anderson and Frost were able to demonstrate this quality in the sera of 66.7 per cent. of a series of abortive instances of poliomyelitis. Anderson and Frost[81] also proved that normal human serum has some neutralizing action but are of the opinion that this has quantitative limits which clearly differentiate it from the serum of the persons who have had poliomyelitis. While such protective bodies do not appear in the serum until two weeks after the onset of the disease, the fact that the virus is known to survive in the throats of experimental animals for much longer periods may make it advisable to test suspected individuals who may be carriers.

Peabody, Draper and Dochez[51] tested out this reaction, taking material from normal persons, from those exposed to infection in the hospital ward and from individuals suspected of having abortive attacks of the disease and from a number of typical cases. The test was performed in the following way: The serum was mixed with a fatal dose of a known active virus, incubated from one to two hours at 37°C. and then allowed to stand 24 hours on ice. The injections were made intracerebrally into monkeys. The protocols of the experiments follow (Macacus rhesus):

Experiment I.—The serum was obtained from a normal adult, one of the physicians in constant attendance on cases of poliomyelitis in the hospital ward. During this time he had presented no signs of illness. One cubic centimeter of the fresh serum was mixed with 0.1 c.c. of virus and was incubated for two hours at 37°C. After standing over night on ice the mixture was injected intracerebrally into a monkey. After 17 days, the animal began to show signs of weakness and was found dead on the following day. At autopsy the characteristic lesions of poliomyelitis were present in the spinal cord.

Experiment VI.—The serum for this experiment was obtained from M. S., two and one-half years old, a patient in the hospital suffering from cerebral hemorrhage. Two tests were made with this serum; in one, 1 c.c. of serum was mixed with 0.1 c.c. of virus and in the other 1 c.c. of serum with 0.3 c.c. of virus. The first animal died of poliomyelitis in seven days and the second in eight days.

Experiment VIII.—The serum for this test was obtained from Donald P., the brother of a patient who died of poliomyelitis in the hospital ward. He had shown suspicious symptoms at the time of the other child's illness. The serum was obtained about three weeks after the suspected attack and 1 c.c. of it was mixed with 0.1 c.c. of virus and injected intracerebrally into a monkey. Ten days later, the animal died, having developed a characteristic poliomyelitis.

Experiment XIII.—The serum used for this experiment was obtained from M. G., four years old, a patient in the poliomyelitis ward. On admission there was a partial paralysis of the external ocular muscles, high fever and some stiffness of the neck. The maximum cell count in the spinal fluid was 320 cells per centimeter. One cubic centimeter of serum obtained 30 days after the onset of the disease was mixed with 0.1 c.c. of active virus and injected intracerebrally into a monkey. Protection was complete, the animal showing no evidence of poliomyelitis.

From these results, it seems that in typical instances of poliomyelitis, the serum of individuals having recovered from acute poliomyelitis always contains after a certain interval protective bodies and that these symptoms persist for a very long time. The sera of the six normal individuals tested, two failed to protect animals against infection whereas four gave complete protection. The sera of suspected individuals, of three tested, one protected, and two failed to protect, while the four sera from individuals who had characteristic poliomyelitis protected animals from fatal infection in every instance; in one case, 30 years after the occurrence of the disease.

Notwithstanding these irregular results one cannot rule out the specificity of the test in apparently normal persons whose serum protects; the possibility must always be borne in mind that he may previously have suffered from an unrecognized abortive attack of the disease. The accurate determination of the protective value of normal serum can probably be made only by a study of sera in very young individuals in whom the possibility of previous infection can certainly be excluded.

Bacteriology.—Throughout the literature one finds many references to various organisms having been found in specimens from this disease. Goldscheider,[82] in 1893, obtained negative results by using methylene blue and Gram's stain in the study of material and failed to find any bacteria in the cord. Siemerling[82] and Dauber used well-recognized bacterial stains and were unable to get positive results. However, Schultze in 1898 described a case in which from the spinal fluid he obtained "many diplococci, arranged in rows, similar to gonococci but having the appearance of the meningococcus of Weichselbaum and Jäger." He, however, failed to cultivate these organisms, though they were numerous. Following this report of

Schultze, Bülow-Hansen and Harbitz[82] in 1898 found a Gram-positive diplococcus in the spinal fluid. They did not attach much importance to it because of their other very numerous negative results. Courmont and Bonne in 1899, Concetti in 1900, Dercum in 1900, Looft and Dethloff in 1901, Gossage in 1902, Engel in 1900, Spiller in 1903, Batten in 1904, and Barns and Miller in 1907,[82] all found microörganisms which they thought to be in some cases meningococci, in others pneumococci or staphylococci. During the Norwegian epidemic of 1905, Giersvold[82] found Gram-positive cocci shaped like beans in the spinal fluid of 12 cases. He inoculated animals with the germ and produced paralysis and death. Harbitz and Scheel[82] also found during this epidemic organisms which grew on ordinary culture media. They, however, reserved judgment on the exact etiologic significance of these organisms. Harbitz and Scheel speak of certain micrococci which they found in three cases in the spinal fluid. These diplococci were bean shaped and grew in artificial media in from two to six days. They say that other workers have also found similar diplococci in other cases of poliomyelitis and that the cocci had been able to produce atrophy, paralysis, emaciation and death, in animals inoculated.

Pasteur, Foulerton and MacCormac[83] in 1908 reported a case of acute paralysis associated with diplococcal infection of the spinal sac. They report a micrococcus which they found in the spinal fluid of a non-fatal case and which, on injection into rabbits, produced symptoms resembling the disease in human subjects. The same micrococcus was identified by them in the spinal fluid of the experimental animals. They were, however, unable to reproduce it in culture.

Leiner and v. Weisner[84] have pointed out the source of error in cultures from spinal fluid, in that they were able to obtain growth of cocci fairly readily from the first portion of lumbar puncture, whereas the last of the fluid was sterile. They tried this in other conditions than infantile paralysis and showed the same conditions to exist. From this they took it that these were contaminations, probably from the skin of the patient. They also pointed out the danger of using broth culture because bacteria will flourish in the broth without giving any indication of the number originally present.

Koch's axiom is still more or less justified in this connection, that for proof of the etiologic significance of any organism, it must be shown to be present in a quantity proportionate to the severity of the clinical signs.

Römer's careful examinations, both by bacteriologic methods and by staining, are worthy of consideration and his negative results are important. He says very emphatically that "it was impossible that a microbe which could be stained and cultivated easily could be the active agent in the causation of epidemic paralysis." And so when in 1909, Landsteiner and Popper[85] first described their transmission experiments and proved that the Heine-Medin disease was caused by a filterable virus, there had already risen unanimously in the field of scientific workers, a belief that a bacteriologic cause for the disease did not exist.

In 1913, Flexner and Noguchi[86] published their experimental work on the cultivation of the microörganism causing epidemic poliomyelitis. They were able to grow from poliomyelitis tissue, brain and cord, of typically affected monkeys, a micrococcus which grew anaërobically in a culture of ascites dextrose broth with fresh sterile tissue. The microörganism as described by them, and later by Amoss[87] and other writers of the Rockefeller Institute, appeared as a globoid or small coccus in pairs or short chains. They were able to find this microörganism not only in cultural media but also in the tissue of infected human and experimental material. The organism was rather short-lived but Amoss has been able to carry it through several generations, each time being able to recover it from the experimental monkey. This organism is apparently filterable through a Berkefeld filter as it has been recovered from the filtrate as well as from the crushed tissue.

The organisms which Flexner and Noguchi and later Levaditi[88] described as having been grown by them in serum-bouillon, have not been capable of constant growth by other workers. Thus Römer and Leiner and v. Weisner have not been able to grow the virus in any media. One must be cautious, as the authors (Flexner and Lewis, Noguchi, and Levaditi) themselves were in accepting these small cocci-like bodies as being the cause of Heine-Medin disease.

It was again somewhat of a shock to the scientific world to hear the announcement during the summer of 1916, from a number of workers, that an easily cultivated polymorpho-streptococcus had been recovered fairly constantly from acute cases and which they considered as the etiologic cause of epidemic poliomyelitis.

Rosenow,[89] Herzog,[90] Nuzum[91] and Mathers[92] have grown a polymorphicstreptococcus-like organism under anaërobic conditions such as Noguchi first described for the micrococcus organism of the virus of poliomyelitis. Mathers has been able to carry the organism through four generations. This streptococci organism, as well as Flexner and Noguchi's microörganism,[86] is short-lived under the best of cultural conditions.

These workers, however, have been able to go further than the older bacterial advocates, in that they have apparently produced the disease in monkeys and rabbits by the injection of these short-lived polymorpho-streptococci and Rosenow[89] claims that he has produced a more or less potent serum from horses by repeated injections of the polymorpho-streptococcus.

Mathers'[93] description of his work on the growth of the polymorpho-streptococcus-like organism which he isolated from autopsy material, the brain and the spinal cord, and his technique are as follows: "Small pieces of nervous tissue were washed in sterile, normal salt solution, crushed and planted in ascites-fluid media and ascites dextrose agar containing pieces of fresh sterile rabbit kidney, coagulated normal horse serum, and ascites dextrose broth. The cultures were incubated at 35 to 37°C. under both aërobic and anaërobic conditions for from 1 to 10 days. Usually growth appeared in some of the cultures in from one to five days."

Mathers made cultures from blood and cerebrospinal fluid before death, and of the brain, spinal cord, spinal fluid, blood, liver, spleen and mesenteric lymph glands after death. In four cases of blood cultures and seven cases of cerebrospinal fluid made before death, all remained sterile. Ten cases of poliomyelitis were studied after death. In nine of these cases a peculiar, polymorphic-streptococcus-like organism was obtained from the tissues of the central nervous system and in seven cases, in pure culture. In two cases staphylococci and in another case B. subtiles were associated with this organism in the brain cultures. In five cases, the cerebrospinal fluid was examined after death and the organism isolated in pure culture in every instance. In two cases, the liver, kidney, spleen and lymph-nodes were cultured and the micrococcus was isolated from mesenteric lymph-nodes and kidneys in one case. He did not find the organism (microscopically) in the tissue of the central nervous system of these cases. Cultures of this were used to infect rabbits and produced lesions of various groups of muscles and characteristic lesions in the central nervous system. This micrococcus has been recovered from the lesion in the inoculated rabbits by both cultural and microscopic methods. The artificial culture of the poliomyelitis coccus in an ascites fluid tissue media under anaërobic conditions causes certain changes in the media which cannot be differentiated from those previously described and cultures of the so-called virus of poliomyelitis. Morphologically, also, this bacteria, when grown in the same media, is similar to the virus and in stained smears, it appears in minute Gram-positive coccus-like forms, arranged in pairs, groups and chains. These minute forms disappeared when the organism was cultivated in other media under anaërobic conditions.

However, doubts as to the importance of even these become very great when we compare them with the vast number of negative results obtained by workers in every quarter of the world, Scandinavia, Austria, Germany and France, and this country generally. Negative evidence by all these accumulated workers is overwhelming. The very careful work in this country of the Rockefeller Hospital and Institute is as good evidence that these various findings of cocci must be discounted at least until further proof is had. Whether the specificity of this polymorpho-streptococcus-like organism can be accepted is at present considered rather doubtful by most workers who have studied the problem for any length of time.

Classification of Cases.—A simplified classification is to be desired. No two cases are probably ever alike. There are few diseases that have a more diversified clinical picture than has poliomyelitis. As can be readily understood, a pathologic process which may involve the brain, medulla, pons, spinal cord and spinal ganglia, or which may be localized in any part of this system, can produce signs and symptoms of very diverse character. The most complete classification is that of Wickmann:[94]

1. The spinal poliomyelitic.
2. Cases simulating Landry's paralysis.
3. The bulbar or pontine form.
4. The encephalitic form.
5. The ataxic form.
6. The neuritic form.
7. The meningeal form.
8. The abortive form.

This classification is based on the combined pathologic and clinical aspects of the disease and is a mixture of the two. The anatomic forms are the spinal, bulbar and encephalitic and the symptomatic forms are the neuritic, meningeal and ataxic. It would be more satisfactory perhaps to group them either anatomically or clinically and would avoid any confusion arising from the use of two symptoms. It is impossible to draw any hard and fast lines between some of the groups; thus most neuritic forms are also of the spinal type and most meningeal ones are either spinal or bulbar. Cases of Landry's paralysis are usually of the spinal type in which the process advances to death because it involves the respiratory muscles. The pathology of the meningitic or neuritic cases is not essentially different from that of the bulbar or spinal. Such a dual classification not only causes deflection or overlapping but the picture of the disease is made more complicated and obscure.

Müller[95] has adopted an anatomic basis and divides his cases into four groups or forms:

1. The spinal form.
2. The bulbar form.
3. Cerebral form.
4. The abortive cases.

From a pathologic, physiologic standpoint, cases might better be divided into two main groups:

1. Those cases in which the upper motor neuron is primarily affected (Müller's spinal and bulbar type).

2. The larger group of cases in which the lower motor neuron is involved (Müller's spinal and bulbar forms). The abortive cases form a distinct group by themselves. The pathologic result of involvement of the lower motor neuron lesions may be in the nuclei of the pons, medulla or the anterior horn cells, the result being in either instance a flaccid paralysis with subsequent atrophy.

The upper motor neuron type or the cerebral type is characterized by spastic paralysis or by the evidence of lesions above the lower neuron. The upper neuron may be attacked in either or both of two places, either the cortex with involvement of large areas or the pyramidal tracts either high up or in the cord. That this class of cases originally described by Strümpell as polioencephalitis acute is very uncommon but that it is a form of infantile paralysis has been made quite certain by epidemiologic studies which show the spastic type occurring in the same epidemic and in the same house with the flaccid type, by occasional occurrences of both spastic and flaccid paralysis in the same patient as well as by the pathologic studies of Harbitz and Scheel.

It would be better then to recognize three groups of cases:

1. The abortive case which never becomes paralyzed.

2. The cerebral group which contains the rare cases in which involvement of the upper motor neuron with resulting spastic paralysis is the chief characteristic.

3. The bulbo-spinal group, much larger, which comprises all cases with lesion in the lower motor neuron and flaccid paralysis. Of course most bulbo-spinal cases show some pathologic foci in the brain and the cerebral cases may show lesions extending into the cord and bulbo-spinal cases may show symptoms of ataxia, the results of which may be in the upper motor neuron either in the brain or cerebrum or in the pyramidal tracts or in Clark's columns. It is impossible to make any hard and fast classification which would apply strictly to all cases. It is best to reconcile the chief clinical symptoms with the predominant anatomical lesions.

Emerson[96] has made a classification based wholly on pathologic anatomy:

1. Non-paralytic or abortive type. In this group are the cases which do not show paralysis; also those which though they have had meningitic symptoms do not go on to paralysis. There may be a disturbance of the sensorium giving the picture of encephalitis. This is probably the largest group in any epidemic and may represent from 50 to 80 per cent. of all cases. In the New York series, however, of the cases admitted to the hospitals, only 15 per cent. did not show paralysis at any time.

2. The ataxic type. In this group which is a comparatively small one, to which attention was first called by Wickman, the virus does not involve the motor cells but the cerebrum, the intervertebral ganglia and Clark's column are involved. The symptoms are lack of coördination, ataxia and nystagmus.

3. The cortical type. This group is also a rare one in which the upper motor neurons are affected, spastic paralysis being the result.

4. The ordinary spinal or subcortical type. The lower motor neuron is affected and flaccid paralysis results. This last group includes the spinal, bulbo-spinal, bulbar, ascending or Landry's type of paralysis, the polyneuritic and those cases with meningitic symptoms which go on to paralysis.

Draper[97] has divided the cases into three groups. The first two groups show signs of general systemic infection. In either of these groups recovery may be complete or paralysis may appear following the systemic phase. This systemic phase represents the period in which the virus is lodged in the spleen or bone marrow as has been pointed out by Flexner and Amoss.[98] So far the first two groups are alike. Then one of two courses may occur in those cases which go on to show involvement of the central nervous system. In the first series there is a complete remission which may last for a few hours to a few days and then the symptoms showing involvement of the central nervous system may appear. This Draper calls the "dromedary" group because of the two humps in the period of symptoms.

The second series, instead of having a complete remission or intermission, continues the infection and involvement of the central nervous system makes its appearance. Draper considers that in such cases the two humps are fused, there being no period of well being in between.

The third series is the sudden onset group. In such cases Draper thinks the systemic phase is either of very short duration and is overlooked or that the virus reaches the central nervous system very rapidly. These cases, however, may completely recover or may go on to paralysis. The appearance of meningeal symptoms or central nervous symptoms is simply a stage in the passage of the virus through the barriers of the meninges.

Predisposing Causes.—**Race.**—There is apparently no immunity to the disease by any race nor any racial susceptibility. It has been thought at times that negroes were less likely to get the disease than the white race but this is not shown by the analysis of the cases either in New York[99] or in Baltimore.[100] In the New York epidemic Germans, Austro-Hungarians and Irish were apparently less involved than those born in Russia and Italy or in the United States.

Sex.—Males are more often affected than females in the average proportion of 3:2. There is no explanation of this fact though we know that certain infectious diseases show this variation between sexes. There is apparently a higher mortality in the males than in the females. The election of males over females is shown less in younger children than in adults.

Age.—The disease is one which attacks practically children under 10 years of age. About 90 per cent. of all cases are under this age. However, in various epidemics such as the Norway epidemic of 1868, 14 per cent. were in adults. Again in the Norway epidemic of 1911, 25 per cent. of all cases were adults. In Iowa in 1910 adults were affected in over 12 per cent. of the cases. On the Island of Nauru[101] in 1910 there were 700 cases, the majority of which were in adults. The mortality in adults is also usually higher than in children though this is not invariably true. The following table[102] gives the result of 1325 fatal cases according to age:

AGES IN TERMS OF PERCENTAGE

Under 1 year	13.0 per cent.
1+ year	22.0 per cent.
2+ years	18.0 per cent.
3+ years	14.0 per cent.
4+ years	9.0 per cent.
83 per cent. were 5 years and under	
5+ years	7.0 per cent.
6+ years	5.0 per cent.
7+ years	3.0 per cent.
8+ years	2.0 per cent.
9+ years	1.5 per cent.
15 per cent. were from 6–16 years	
10+ years	0.75 per cent.
11+ years	0.375 per cent.
12+ years	0.375 per cent.
13+ years	0.75 per cent.
14+ years	0.375 per cent.
15+ years	0.375 per cent.
Number cases under 16 years	97.5 per cent.
Over 16 years	2.5 per cent.

Type.—Draper[103] has called attention to the very interesting fact in relation to the type of child which is most affected. This is usually the "large, well grown, plump individual who has certain definite characteristics of face and jaws, is broad-browed and broad and round of face."

The teeth, especially the central incisors of the upper jaw, are widely separated, in fact all the teeth may stand out entirely free. On the other hand, in adolescent and young adults, Draper calls attention to the fact that in these individuals the type is entirely different. They are usually brunettes, with delicate and dark skin, high color of the cheeks and lips. Instead of being robust they are delicate and instead of the teeth being widely spaced they are close together, there being a tendency to crowding and definite maxillary prognathism.

The disease occurs in greater proportion to the population in rural districts than in cities. This is even true within cities as was shown in the New York epidemic where there were proportionally more cases in the boroughs of Queens and Richmond, which had a rate of 3.22 and 2.92 per thousand population respectively as compared with the boroughs of Brooklyn, Bronx and Manhattan, which had a case rate respectively of 2.24, 1.16 and 0.94 per thousand estimated population. The same is shown in the following table:[104]

EPIDEMICS IN SWEDEN

Years	1905	1906	1907	1908	1909	1910	1911
Cases	1016	429	467	317	178	109	3840
Percentage in towns	7	3	23	11	8	6	19

Social and Hygienic Conditions.—Neither the social condition nor the hygienic condition seem to influence the disease as has already been illustrated from the incidence on Barren Island, the dumping ground for the garbage of New York City. The well-to-do were affected as much as those who lived in the tenements. The following table[105] of a thousand cases from the borough of Brooklyn shows that the class of dwelling has little to do with the incidence of the disease:

241 cases occurred in private houses,
12 cases occurred in boarding houses,
747 cases occurred in tenement houses.

Curiously enough Professor Brues' studies[106] brought out the fact that the two lower floors were more often affected than the upper floors as is shown by the following table:

	Age under 1 Year	Age over 5 Years
Basement, first and second floors	49.3	44.8
Third floor and above	50.6	55.2
Number of cases	227.0	301.0

"In this connection it is noticeable that in children under one year old, at which age it is probable that they play more regularly in the apartment in which they live, the divergence between cases occurring on lower floors and upper ones is greater than in older children, who, it may be expected, spend more of their time down stairs or on the street." And the Editor appends this note:[106] "This study is incomplete and the conclusion therefore not fully justified owing to the fact that no count was made of the children of the specially susceptible ages, or the children of all ages living in apartments on the various floors of the tenements. It cannot be safely assumed that there is an equal distribution of children of all age groups on the different floors of the tenements of New York City."

Brues attributes the above finding to the fact that rats do not commonly occur on upper floors of buildings. He also points out that from this standpoint it differs from diphtheria and scarlet fever in its distribution on various floors.[107]

Basement, First and Second Floors	Per Cent.	Third and Higher Floors	Per Cent.
Poliomyelitis	44.3	Poliomyelitis	54.1
Diphtheria	39.9	Diphtheria	58.9
Scarlet fever	37.0	Scarlet fever	60.5

Of this Brues says "this difference implies an infective agent which is more prevalent near the ground; the only other plausible assumption would seem to be that the disease is favored more by the lack of air and light on lower floors than are scarlet fever and diphtheria."

Conditions Existing in the Mouth and Throat.—By some it has been suggested that teething has a relation to poliomyelitis. This is an interesting observation and has been referred to by Draper and others as at least being suggestive. The fact that poliomyelitis occurs more often during the period of teething, however, may or may not have anything to do with susceptibility. As has been pointed out by many workers it may simply refer to the age period during which previous infection of the disease has not occurred. In the same way this is true of almost all contagious diseases which are more prevalent during childhood than during adult life, as measles and whooping-cough. No one would think of referring these to any question of teething, most adults having had these diseases or not having had close enough contact to develop them. Decayed teeth have been supposed by some to be the channel through which infection might enter. The tonsils and adenoids or the mucous membrane of the naso-pharynx is undoubtedly more often the route of entry. This was studied in the New York epidemic where it was found in a small group of cases in which the tonsils and adenoids had been removed that 46 per cent. of these cases recovered whereas only 15 per cent. of those who had not had their tonsils and adenoids removed recovered. It has been shown by almost all workers that the tonsils and adenoids will harbor infection for long periods. The virus may be obtained from the nasal secretions as long as four months after the acute stage. In one case reported by Lucas and Osgood[74] these washings were positive 204 days after the acute stage. It would appear from the few studies that have been undertaken that children who have had their tonsils removed are less susceptible than those who have not.

Symptomatology.—Incubation Period.—From experimental studies it would appear that the incubation period varied from 3 to 10 days, the average occurrence being from five to seven days. Cases have been reported as early as 24 hours after contact and as late as 16 or 18 days.

Symptoms of Systemic Infection.—Symptoms of the systemic period may be those of simple general infection. First, fever-temperature is usually present at the onset at least in the epidemic cases. Temperature usually ranges between 101 and 103°F., though in certain of the very acute cases temperature may run as high as 104 or 105°F. Temperature usually comes down by crisis within a few days though many cases may run a temperature from 99 to 100°F. for several weeks, often into the period of convalescence. Wilson[108] in a study of 400 cases in the Willard Parker Hospital found fever absent in only 2 per cent. of cases and it was the initial symptom in 334 cases out of 400.

At times the first symptom is that of lassitude or drowsiness. The onset may almost be that of coma though usually these periods are interrupted by periods of restlessness and irritability. There may be no apathy or mental dullness and the patients may be very bright and mentally active. Wilson[108] found persistent drowsiness in 72 per cent. of his cases.

There is usually a loss of appetite accompanying the fever. The pulse rate and the temperature usually follow the same course, the pulse is not generally markedly accelerated except in very severe cases. In some cases meningeal irritation may be very prominent. Convulsions may be present from the onset. Not all these cases go on to paralysis. The condition may clear up entirely so that the severe onset does not necessarily foretell a severe involvement of the central nervous system. The gastro-intestinal system is usually affected. In young infants this may be shown by vomiting or by diarrhea though constipation is more often present than diarrhea. The constipation may continue for a long time, especially if the abdominal muscles are affected.

Some of the cases are ushered in with a general pain or sensitiveness; this may be in the form of a general hyperesthesia, so that the slightest touch may cause pain or the pain may only be elicited on deep muscle pressure or on movement of any part of the body. Pain is most frequent along the spine. At times there may be a fine tremor of the hands or jerky movements of any part of the body. At times the part later paralyzed is the one that shows most of these jerky movements. In older cases severe headache, especially frontal headache, is complained of. One of the characteristic symptoms is the elicitation of pain when the neck is flexed forward. This is considered by some a very important sign. It rarely goes on to opisthotonous though the head is often held slightly flexed backward. Kernig's sign, however, is not usually present.

There is often a marked degree of sweating. This is generally only present during the febrile period but it may continue into convalescence. It is more often found when there are muscle tremors and jerking movements and it is supposed to be due to involvement of the cells which govern the sweat function.

During the early stages in some cases there may be an acute sore throat. As has been pointed out by Draper[109] this systemic phase may be followed by a period of well-being (dromedary group) or the systemic phase may go directly into the paralytic. There is no symptom outside the appearance of paralysis itself that indicates the onset of paralysis. The following table from the New York Epidemic[110] showed that the paralysis occurred in 71 per cent. of 700 cases before the fourth day:

189 had paralysis on the first day of illness
203 had paralysis on the second day of illness
142 had paralysis on the third day of illness
85 had paralysis on the fourth day of illness
48 had paralysis on the fifth day of illness
29 had paralysis on the sixth day of illness
20 had paralysis on the seventh day of illness
14 had paralysis on the eighth day of illness
6 had paralysis on the ninth day of illness
3 had paralysis on the tenth day of illness
2 had paralysis on the eleventh day of illness
2 had paralysis on the twelfth day of illness
1 had paralysis on the thirteenth day of illness
4 had paralysis on the fourteenth day of illness
1 each on the fifteenth, sixteenth, twentieth and twenty-first days.

The paralysis may be perfectly evident or it may only be found after very careful searching. In very young children it is often difficult to make out whether paralysis is present during the acute stage or when it begins. In older children it is comparatively easy. Paralysis in young infants may be brought out by gently pricking the parts supposed to be paralyzed. Very often it is difficult to tell whether the child does not move the limb because it is paralyzed or because of pain or fear. Often testing out the resistance of a group of muscles by flexing the limb and then suddenly straightening it out may assist in determining whether paralysis is present; normally there will be a definite muscle resistance to the sudden stretching. It is, however, hard to define whether the lack of resistance is due to paralysis or to general weakness. The paralysis may be limited to a group of muscles, to a single muscle or it may be generalized. The paralysis of a single group may not even be complete. It may be partial paralysis or only a temporary weakness. Where the paralysis is general or where it affects a large muscle group it is comparatively easy to make the diagnosis. Where it is partial it may offer considerable difficulty. For these cases Lovett and Martin[111] have proposed a spring balance test by which any muscle or group of muscles can be measured. Lovett[112] feels that the distribution of the paralysis is much more general than has been usually supposed. Even in the cases in which there is no apparent paralysis they have found a considerable number of muscles by these tests which show definite signs of partial paralysis. They estimated that the proportion of partial paralysis to total paralysis of muscles is as 9:1. This test, however, is rarely applicable during the acute stage but is of great value during the convalescent and after period. The distribution of the paralysis varies considerably, more often one leg is paralyzed, next in frequency both legs. The following table gives the distribution of the paralysis:

DISTRIBUTION OF PARALYSIS (1907–1910). REPORT OF THE MASSACHUSETTS[113] EPIDEMIC OF 1910

	Cases	Per Cent.
One leg only	324	27.97
Both legs only	272	23.48
Back	154	13.29
Both arms and both legs	129	11.13
One arm and leg, same side	110	9.49
One arm only	84	7.25
Both legs and one arm	75	6.47
Face	74	6.38
Abdomen	67	5.78
One arm and leg, opposite sides	33	2.84
Respiration	31	2.67
Both arms only	23	1.98
Neck	11	0.94
Both arms and one leg	10	0.86
Deglutition	7	0.604
Neck and back	6	0.51
Ataxia (transitory)	5	0.43
General	3	0.25
Intercostal	1	0.086
Both arms, back, chest and throat	1	0.086
Total	1420	

The Monograph of the Department of Health[114] also gives the following table as to the incidence of paralysis in 1065 cases in the Willard Parker Hospital:

Involvement of lower extremities only	529
Involvement of upper extremities only	104
Involvement of head and neck	122
Involvement of lower and upper extremities	111
Involvement of back and lower extremities	139
Involvement of back and upper extremities	12
Involvement of face with upper extremities	7
Involvement of face with lower extremities	19
Involvement of back only	20

Anatomical Considerations.—The spinal motor cells lie in spindle-shaped bundles in the anterior horns of the spinal cord, being chiefly located in the cervical and lumbar enlargements. These bundles lie longitudinally, reaching through two or three spinal segments, the various groups overlapping one another. This accounts for the irregular and complex involvement of muscles which one sees, as a single center will send impulses to two or more muscles and one muscle will receive motor impulses from more than one group of cells. On the other hand, the blood supply, which is derived chiefly from the anterior and two posterior spinal arteries, runs horizontally and as the virus reaches the cells through the blood supply it is easily understood how partial paralysis is more frequent than complete; furthermore the arteries are terminal ones and do not anastomose, so a single chain of cells may be involved which may supply motor impulses to several muscles but not completely to any one.

Spinal Form.—As the anterior horn cells of the cord are the ones most often affected this form is the one most often seen. Any group of muscles may be affected. In the leg the quadriceps femoris is the one usually paralyzed while in the lower leg the anterior group of perineals, which are the flexors of the foot and extensors of the toes, are most often paralyzed. The paralysis is usually more extensive at first; it may clear up partially or completely. The flexors of the toes are rarely affected and for this reason the most common paralysis produces drop foot.

Lovett points out that the hip is more frequently inolved but the completeness of the paralysis is more in the lower leg. Both legs will be affected about equally. On the other hand, the left arm is more often paralyzed than the right and as a rule only one arm is paralyzed. When the deltoid is affected this is usually more extreme than paralysis of the arm. Lovett thinks that the frequency of paralysis is in proportion to the function of the muscles involved. The more complicated and continuous are the movements of any set of muscles the less likely are they to be involved. For this reason the left arm is often more involved according to Lovett than the right. This may be due to the fact that on account of its greater activity there is a greater blood supply. There is another factor in the question of weight bearing.

The muscles that have the greatest amount of weight to bear are more often affected. Thus the shoulder muscles are more frequently affected as they carry most of the weight, and the lower leg muscles more often than the hip because the greater weight falls on them. Therefore, according to Lovett the anatomical distribution of the blood supply as well as the functions of the muscles and their activity has something to do with the amount of complete or partial paralysis.

Paralysis of the back muscles is very common. In young children it is often difficult to make this out during the acute stage as they are usually unable to sit up. These muscles are seldom affected alone, usually one of the extremities will be affected at the same time. Fortunately paralysis of the back muscles clears up in part at least and rarely remains complete and then usually only unilaterally, producing a scoliosis.

Paralysis of the neck muscles can be made out by lifting the child by the shoulders, if the head falls back and the child is unable to raise it, there is good evidence for believing that paralysis is present. It must not be taken for granted that because the child lies with its head extended that it is paralyzed because very often they assume this position as it is more comfortable. Total or permanent paralysis of the neck muscles is uncommon. Where it does occur it is usually associated with very extreme general paralysis.

Paralysis of the diaphragm is often involved in fatal cases. The diaphragm is supplied by the third, fourth and fifth cervical segment through the phrenic nerve. It can be made out by holding the thorax firm which causes labored and difficult breathing without any diaphragmatic effort being seen. Normally if the chest is held firm the abdomen is seen pushed forward, whereas if the diaphragm is paralyzed there is abdominal retraction on inspiration. In the same way the intercostal muscles may be paralyzed; by holding the abdomen the thorax does not move on inspiration, or it may be moved downward instead of upward and forward as it does normally. Both diaphragmatic and intercostal paralyses occur most often in the fatal cases and they are usually late in making their appearance.

Abdominal muscle paralysis is not common. The rectus usually escapes and only a portion of the transversalis, either external or internal, is affected. This can be made out when the child cries, the paralyzed section will bulge definitely.

The cranial nerves are often involved, especially the seventh or facial. This is easily made out by the asymmetry of the face when the child cries. It can often be detected when the face is at rest by the lack of wrinkles, and the smoothness on the paralyzed side and because the eye on the paralyzed side is open. The patient is unable to wrinkle the forehead or nose on the paralyzed side. The corners of the mouth are drawn up on the unaffected side and saliva often drooles from the paralyzed side. Involvement of the seventh nerve is often associated with paralysis of the sixth nerve or external rectus. When this is affected the patient is unable to turn the eye outward beyond the mid line. The third and fourth nerves are less commonly involved. When the third is paralyzed the pupil is dilated and does not react to light. The fourth or superior oblique is only occasionally affected. When the fifth nerve, the trigeminal, is involved, one or all three parts may be affected; it is partly motor and partly sensory. When it is involved food collects on the affected side and there may be a pseudo-facial paralysis due to the involvement of the sensory fibers. The zygoma usually stands out on the affected side due to the atrophy of the muscles. The first and second nerves are rarely if ever affected though the second nerve has been reported as affected with the result of optic neuritis and blindness. The eighth nerve to the ear is also rarely if ever affected. Along with the facial nerve, occasionally there is involvement of the twelfth or hypoglossal nerve. This is easily made out if the tongue is protruded; there is usually marked atrophy of the affected part. The most serious cranial nerve involvement is that of the ninth, tenth and eleventh. When the vagus or pneumogastric nerve is involved there is marked change in the type of respiration. Respiratory distress and rapid heart action are the symptoms; with the involvement of the ninth nerve there is difficulty with swallowing.

When the pons and medulla are involved the cases are spoken of as bulbar paralysis. It is usually associated with spinal cord involvement but may be the only sign of poliomyelitis and is often accompanied by cranial nerve involvement also. All types of combinations may be present. The patients are usually clear mentally, over anxious and their whole expression is very characteristic. If one has ever seen the alertness and expression of anxiety on these patients it can never be forgotten. Mortality in the bulbar type is usually very high as it occurs mainly in the severe type of infection.

Ataxic Form.—This form is rather rare. It was first described by Medin, as already pointed out. The lesions may be either in the brain, cerebellum, pyramidal tracts or Clark's column. It is shown by the staggering gait on walking. The child falls easily, the legs are held wide apart and the disturbance in equilibrium is easily made out. Fortunately this condition is usually of short duration and is not associated with high mortality. In certain cases involvement of the cervical cord with transient ataxia may be present. It is supposed that when the pyramidal tracts or Clark's column are involved in these cases there is a tendency to hold the legs stiff and there is increased knee jerks.

Cerebral Type.—This was first described by Strümpell, as already noted. It is an uncommon type though it has been reported in several of the epidemics. The onset does not vary from the ordinary type though it may be associated with convulsions, but not all cases beginning with convulsions belong to the cerebral type. The resulting paralysis in these cases is of the spastic type. There is no atrophy, no reaction of degeneration and the reflexes are exaggerated. Some of these cases later develop epilepsy or mental derangements. Athetosis of the hands frequently results. A few cases have been reported in which the cerebral type was combined with the spinal type and in which there was both spastic and flaccid paralysis. Instances have been reported in which in the same family one case would have cerebral involvement and the other spinal. Frost and Anderson[81] have reported an instance in which a case started with flaccid paralysis and later became spastic. The lesions in the autopsies which have been described show a rather scattered involvement of the cerebrum.

Meningeal Type.—Meningeal symptoms or meningeal irritation are often a sign of acute poliomyelitis. This may be such a prominent symptom that epidemic meningitis is first considered. Usually, however, the symptoms of meningeal irritation do not last unless there is associated with it further involvement of the pons or medulla.

Polyneuritic Type.—In this type the main symptom is pain and it is often mistaken for a multiple neuritis. It may be associated with very little paralysis.

Ascending Type or Landry's Paralysis.—This was first described by Landry[115] in 1859. Since that time this type of paralysis has been accepted by most workers as a special type of acute poliomyelitis. In epidemics it is not an uncommon form of disease and belongs to the bulbo-spinal group. It may be ascending or descending; in either case involvement of the respiratory centers is common. It is usually associated with difficulty in swallowing and of articulation. The paralysis is generally progressive though it often has a step-like progression. When it is descending the patient may die from respiratory failure before the lower extremities are involved. The mind usually remains clear though in some cases it may be stuporous from the start. It must be differentiated from toxic ascending myelitis due to bacterial or other toxins or from a toxic polyneuritis.

Diagnosis.—In the early acute stage it may often present many difficulties. The chief aid in diagnosis is that of the spinal fluid. The spinal fluid, though it is not pathognomonic, shows an increase in amount and is under pressure. It is usually clear at first though it may have an appearance of ground glass. This may be due to the increased amount of cells present though it may be due to a small amount of blood. A small fibrin web may form though this is not as large as that found in tuberculous meningitis. The cells, the albumin, and globulin are always increased. The cell count may run very high, over 1000 per centimeter, though usually early in the disease it is not increased much over 100 per centimeter. The number of cells is of no prognostic value. The cells can either be counted in the ordinary Thoma-Zeiss counting chamber or may be sedimented and stained. The cells are generally mononuclears though at times polymorphonuclears may predominate. Neal[116] found only three cases in 400 fluids in which the polymorphonuclear cells predominated. These occurred on the third, fourth and fifth days respectively. She thinks that polymorphonuclears when present represent a special type of the disease and are not characteristic of the stage of the disease. Large mononuclears and endothelial cells are more frequent in poliomyelitis than in any other condition. The albumin and globulin are usually increased moderately and their presence persists after the cells have returned to normal. Fehling's is always well reduced. The cell count is usually more evident during the first week and tends to return to normal during the second week. They rarely are increased over 300 or 400. It is important to do lumbar punctures in suspected cases frequently, as often as every 8 or 12 hours, because as will be pointed out in treatment the earlier diagnosis is made the more hope there is of effecting a cure by specific serum treatment. If the diagnosis is delayed until paralysis appears, the hope of effecting any results from specific treatment are definitely diminished if not entirely lost. Following is a differential table of the ordinary findings in spinal fluid in various conditions:

CEREBROSPINAL FLUID IN VARIOUS MENINGEAL CONDITIONS IN CHILDREN

	Normal	Meningismus	Serous meningitis and encephalitis	Poliomyelitis, polio-encephalitis
Color	Clear	Clear	Clear or opalescent	Clear or opalescent
Pressure	Comes in drops	Increased	Increased	Increased
Quantity	Up to 10 c.c.	Increased	Increased	Increased
Clot formation	None	None to very slight	Early fibrin web	Early fibrin web
Fehling's	Reduced	Reduced	Reduction decreased	Reduction decreased
Nonne albumin	No increase	Usually no increase or very slight	Increase slight to 3+	Increased especially late
Noguchi globulin	No increase	Usually no increase or very slight	Increase slight to 3+	Increased especially late
Potassium permanganate.	Index below 2	Around 2	Early 2+ Late 2−	Early 2.5+
Bacteriology	None	None	None	None
Fixation (biologic)	None	None	None	None
Cytology number	10–12	20	50–100+	Early, very high, 1000
Polys			Occas. 50%	Early 60%
Lymphocytes small monos		80–100%	80–100%	80–100%
Large monos				Often high
Endothelial	Few	Few		Numerous

CEREBROSPINAL FLUID IN VARIOUS MENINGEAL CONDITIONS IN CHILDREN.—(*Continued*)

	Tuberculous meningitis	Pyogenic cerebrospinal meningitis	Lues, epilepsy
Color	Clear to opalescent	Cloudy turbid	Clear
Pressure	Increased	Increased	Increased or normal
Quantity	Increased	Increased	Increased
Clot formation	Fibrin web froin coagulation	Clot	None
Fehling's	Tends not to reduce	Decreased or not reduced	Reduced
Nonne albumin	Increased	Increased	May be either
Noguchi globulin	Increased	Increased	May be either
Potassium permanganate	2.5 up to 5+	3+	
Bacteriology	T.B.	Specific	None
Fixation (biologic)			Wassermann
Cytology number	Late high 300+	High	10
Polys	Acute high %	80–100%	
Lymphocytes small monos	Usually 90–100	Few	Few
Large monos	Small %	Few	
Endothelial		Occasional	Few

The conditions which give the most difficulty from the study of the spinal fluid are tuberculous meningitis and so-called meningismus or encephalitis. In tuberculous meningitis the history and onset are entirely definite and in our experience the tubercle bacilli can always be found if enough care and patience are taken in examining the fluids. There is no definite way of differentiating an encephalitis or meningismus from poliomyelitis by examination of the spinal fluid. In epidemics it is always safe to assume that these cases are cases of poliomyelitis. Unless they are associated with some known condition such as whooping-cough, the spinal fluid in cases of prolonged and severe convulsions may resemble that of poliomyelitis and spinal fluid taken just prior to death also frequently resembles that of poliomyelitis. The spinal fluid of acute syphilitic meningitis can be differentiated by the Wassermann reaction. Occasionally in poliomyelitis spinal fluid where there is a true hemorrhagic condition present it is difficult to make any positive statement. Where blood is present it can easily be differentiated from faulty technique because where a vessel has been punctured the first spinal fluid will show evidences of blood, whereas later tubes will be much less cloudy. Where there has been an old hemorrhage at times the fluid will have a characteristic yellow color and will coagulate spontaneously. This type of fluid is occasionally met with but it is also found in other conditions as tuberculous meningitis. The Lange's colloidal gold test is in the luetic series so that this is of no diagnostic aid.

The Blood.—The blood in acute poliomyelitis does not differ from that of any acute infection. There is usually a slight increase in the polymorphonuclear cells. The neutralization test which consists in mixing the blood serum from a suspected case of poliomyelitis with the known active virus, incubating it and inoculating a monkey intracerebrally and using a control monkey in which active virus alone is injected. The control monkey will become paralyzed while the other will not. This test has been used but it is of no value in making a rapid differential diagnosis as it takes from three to seven days ordinarily for the control monkey to become paralyzed. And there is a further objection from the fact that it is impossible to say how many people are immune or have suffered from a previous attack without paralysis. Of course, this does not hold for children as much as it does for adults. Blood chemistry is normal; there is no change in the non-protein nitrogen fractions.

There is nothing in the urine that is of any value. There may be a slight amount of albumin present which is characteristic of any acute infection. There may be retention of the urine in severe cases. The sphincter control is not usually lost though where there is abdominal paralysis retention may be a symptom.

Differential Diagnosis.—The most important point in differential diagnosis is the history. Acute poliomyelitis differs from most of the conditions with which it might be confused, as the pseudoparalyses of rickets or scurvy; in both of these conditions there are other signs which are definite and the history of the cases is entirely different. In tetany the characteristic position of the hands and feet and the reaction of the muscles make for easy differentiation. Differentiation from the various acute involvements of the meninges is usually not difficult after examination of the spinal fluid.

Prognosis.—It is very important not to make any definite prognosis early in poliomyelitis as it is impossible to say during the acute stage what the outcome will be. A case which may begin very mildly may develop rapid ascending paralysis and end fatally, whereas a case which begins with convulsions may in a day or two subside with little or no paralysis. The mortality in the various epidemics has varied very widely, ranging from 8 per cent. in the Massachusetts epidemics of 1910 to over 26 per cent. in the New York epidemic of 1916. Mortality by ages is shown in the following table from the New York[117] series:

POLIOMYELITIS—CASES AND DEATHS

(Rate per 1000 Estimated Population at Each Age Group, June 1 to Nov. 1, 1916)

	Cases			Deaths			Case fatality, both sexes
	Total	Males	Females	Total	Males	Females	
Total all ages	8,914	5,061	3,853	2,406	1,408	998	
Rate per 1000 population	3.80	4.31	3.28	1.03	1.20	0.85	26.99
Under 1 year	982	533	449	423	240	183	
Rate per 1000 population	18.40	19.70	17.06	7.93	8.87	6.95	43.08
1 to 2 years	1,775	1,011	764	521	312	209	
Rate per 1000 population	34.24	38.70	29.71	10.04	11.94	8.13	29.35
2 to 3 years	2,003	1,173	830	435	255	180	
Rate per 1000 population	41.01	47.52	34.48	8.91	10.33	7.45	21.72
3 to 4 years	1,444	820	624	311	185	126	
Rate per 1000 population	29.85	33.50	26.10	6.43	7.56	5.16	21.53
4 to 5 years	852	496	356	207	136	71	
Rate per 1000 population	18.04	20.77	14.65	4.38	5.70	3.04	24.30
Total under 5 years	7,056	4,033	3,023	1,897	1,128	769	
Rate per 1000 population	28.26	31.96	24.47	7.60	8.94	6.23	26.89
5 to 6 years	593	334	259	150	89	61	
Rate per 1000 population	13.23	14.86	11.59	3.35	3.96	2.73	25.30
6 to 7 years	345	202	143	92	45	47	
Rate per 1000 population	7.73	9.06	6.39	2.06	2.02	2.10	26.67
7 to 8 years	226	115	111	67	38	29	
Rate per 1000 population	5.15	5.24	5.07	1.53	1.73	1.32	29.65
8 to 9 years	157	81	76	36	18	18	
Rate per 1000 population	3.72	3.87	3.47	0.85	0.86	0.84	22.93
9 to 10 years	111	62	49	32	18	14	
Rate per 1000 population	2.70	2.99	2.40	0.78	0.87	0.69	28.83
Total under 10 years	8,488	4,827	3,661	2,274	1,336	938	
Rate per 1000 population	18.20	20.58	15.79	4.88	5.70	4.05	26.79
Total 10 years and over	426	234	192	132	73	59	
Rate per 1000 population	0.23	0.25	0.20	0.07	0.08	0.06	30.99

CASES AND DEATHS IN PATIENTS UNDER ONE YEAR
(June 1 to Nov. 1, 1916)

Age in months	Cases	Deaths
Under 1 month	7	3
1 month	19	13
2 months	27	10
3 months	52	17
4 months	70	24
5 months	64	34
6 months	98	45
7 months	118	45
8 months	104	44
9 months	153	52
10 months	149	73
11 months	121	63
Total	982	423

In the New York series 81+ per cent. died during the first week, the greatest number of deaths being on the third, fourth and fifth days; 11+ per cent. died during the second week and 3+ per cent. after the twenty-first day.

Prognosis as to recovery from paralysis is a very difficult thing to make. In general it is fairly safe to predict that there will be a certain amount of recovery and with modern treatment a great deal more recovery is obtained today than was thought possible a decade ago. The various epidemics also vary considerably in the amount of recovery of paralyzed muscles. Wickman reports as high as 44 per cent. as cured; whereas in the Massachusetts epidemic of 1910 only 16.7 per cent. were reported as complete recoveries. In the New York epidemic out of 1223 cases treated by the New York City Department of Health 32.6 per cent. showed total recoveries, whereas 67.4 per cent. showed some paralysis on discharge from the hospitals. The question of functional recovery rests upon how carefully treatment is carried out. Lovett and Martin[119] show that the chances of functional recovery are as 6:1 when the cases are followed by proper muscle training under the guidance of an expert. Where the training is done at home the improvement is as 3.5:1 and in cases in which there was no supervision it was as 2.8:1. Where muscle training was carried out for two months there was improvement in 48 per cent. of cases whereas when there was no treatment there was only improvement in 27 per cent.

Treatment.—Treatment must be divided into that for the acute stage, that for the convalescent stage and that for the chronic stage. The only specific treatment so far suggested is that of the use of serum from patients who have already had the disease. Netter[120] has suggested the use of blood serum from recovered patients and used it in a small series of cases: The rationale of this procedure is based on the fact that, as Flexner and Lewis have shown, monkeys that had recovered from the disease could not be re-infected, and further that the blood of such monkeys had the power of neutralizing virus. This neutralization test has already been referred to as a means of diagnosing suspected cases. The blood apparently has this antigenic property developed very shortly after the acute stage is over and it lasts at least 30 years. A certain amount of immunity is shown to be present in normal human serum and some have even used normal horse serum. Flexner and Lewis[80] showed that the injection of immune serum as late as 24 hours after the monkey had been inoculated with active virus would prevent the disease or at least produce a very mild type as compared with the control monkey injected at the same time with the same amount of active virus. Amoss and Chesney[121] treated a small series of cases (26) and found that 71 per cent. of these did not show any paralysis. They treated their cases both by injecting the serum, intraspinally and intravenously. This method was advocated by the New York Health Department. They recommended 15 c.c. of serum to be injected intraspinally, after the removal of a slightly larger amount of spinal fluid, by the gravity method, the tests to be repeated every 20 to 24 hours, two or three injections to be given. Where there is rapid progress of the condition injections may be given as often as every 12 hours. In some cases there was a rise of temperature and a change in the spinal fluid which showed a marked increase in the polymorphonuclear cells.

The following is taken from the New York report:[122]

"**Method of Obtaining and Preparing the Serum.**—(*a*) *Amount.*—To obtain the immune serum the blood is drawn from suitable donors in quantities varying with the age, weight and apparent hemoglobin content of the individual. On an average, it is safe to withdraw 2 oz. from children 9 to 10 years of age, 3 to 4 oz. from children 12 to 13 years of age, 4 to 6 oz. from individuals 18 years of age and over. Adults, especially robust, full-blooded persons, can furnish 10 to 16 oz. of blood. Similar amounts of blood can be safely withdrawn again at the end of two or three weeks.

"(*b*) *Technique of Obtaining the Blood.*—The blood is obtained either by means of a No. 15 gage steel or platinum needle, to which a small piece of rubber tubing is attached. In children, adults and stout individuals with small or indistinct veins, the blood is withdrawn by means of a 1 oz. Record syringe and a No. 17 gage needle. The blood is collected in small square bottles in quantities of 1 to 2 oz., and given a long slant so as to obtain as broad a surface for the separation of the serum as is possible.

"(*c*) *Preparation of the Serum.*—The blood is allowed to clot, and the bottles are then placed in the ice-box during the following 24 hours to allow a separation of the serum. This is decanted the following day and centrifuged to free it from pieces of blood clot and red blood cells. To the serum is next added a preservative in the form of 0.2 per cent. trikresol. (The trikresol increases the local irritant action of the serum, and it may be found advisable not to add it as a preservative.) This is added in a 25 per cent. solution in quantities of 4 c.c. to every 500 c.c. of serum. The serum is then allowed to remain in the ice-box for 48 hours, so that a fine precipitate, which forms after the addition of the trikresol, separates out and is removed. The serum is then passed through a Berkefeld stone filter, either by suction or pressure, bottled in quantities of 15 c.c. in dark-amber or blue bottles, and kept cold in the ice-box.

"(*d*) *Duration of Efficiency of the Serum.*—If the serum has been preserved with trikresol, or handled with sterile precautions after it has been passed through the Berkefeld filter, and is afterward kept in a cold place, it will probably remain efficient in its specific content for a number of weeks. The serum obtained was used up almost as fast as it was obtained. Some of the serum which was kept for four to six weeks seemed to be as active, therapeutically, as the more recently drawn serum.

"In an emergency, or where the facilities for treating the serum are not obtainable, the blood is simply drawn under aseptic conditions in a vessel with glass beads and shaken and centrifuged; or the serum, drawn in the usual way, is allowed to separate during the next few hours, and promptly used, disregarding entirely the presence of the few suspended red blood cells."

They found that where the serum was injected early before the paralysis occurred there was an increase in the meningeal symptoms, increased rigidity of the neck, opisthotonos, Kernig's hyperirritability, headache, vomiting, increased temperature which at times reached as high as 104 or 105°F., convulsive movements or twitchings may be seen. In reviewing their results they state that it is "Interesting to see that of 54 preparalytic cases treated with immune serum, 44 remained free from paralysis, while of the 10 who developed some form of paralysis five made a complete final recovery."

Their results were not nearly so good in cases treated after paralysis had occurred. Of the 102 cases that lived 12 recovered completely and 90 showed improvement at the time they were discharged from the hospitals. They summarized their results as follows:[123]

"*Summary of Results.*—It is known that in poliomyelitis we have a group of non-paralytic or abortive cases which go through the premonitory symptoms, but do not terminate in paralysis. It is impossible to state, therefore, how many of the cases treated with serum would have remained free from paralysis without serum treatment. The results and the conclusions from any form of treatment in a disease which is so varied, variable both in symptomatology and in prognosis, as to life and as to function, must be given with reserve.

"The results obtained seemed to be favorable when the serum was used in suitable cases in the pre-paralytic stage of the disease. Paralysis has been followed in some cases, but the mortality seems to have been influenced by the administration of serum.

"The later and more severe cases treated after the paralysis had already made a distinct headway and was beginning to involve the muscles of respiration, showed, in a certain proportion of cases, an inhibitory effect of the serum upon further progress and a possible life-saving result.

"While no absolute judgment of the value of a serum can be based as yet upon the results obtained, they are, nevertheless, encouraging and justify a continuation of the serum treatment in acute poliomyelitis until, in the course of time, more definite data be available."

Further, after discussing the experimental value of this serum, they say:[124]

"In the minds of the general public and of most physicians, there exists a close analogy between the use of serum in epidemic meningitis and in poliomyelitis, and its success in the former has influenced them to have faith in the success of the latter. The pathology of the two conditions, however, is quite different. In acute purulent meningitis the process is limited almost entirely to the meninges, the brain and cord substance being little if at all involved, though there may be some secondary congestions. In poliomyelitis the pathologic picture is reversed. Here the brain and cord substance is mainly involved, whereas the inflammation in the meninges is entirely secondary. Injecting a foreign substance into the slightly inflamed meninges sets up in most cases an acute aseptic meningitis, as is shown by changes in the spinal fluid, and clinically, by increased temperature, rigidity of the neck and other signs of meningeal irritation. It is reasonable that this increased inflammatory reaction should tend to accentuate the inflammatory changes existing in the subjacent substance of the brain and cord. This is contrary to the commonly accepted idea that the first indication in the treatment of an inflammation is rest. And that this reaction may be harmful is borne out by the results of animal experimentation. In meningitis, on the other hand, the injection of serum into actively inflamed meninges is not followed by an increase in the inflammatory reaction, as is evidenced by the clearing up of the fluid and the amelioration of the clinical symptoms in favorable cases.

"It would seem, therefore, that from this point of view the action of the serum may even be distinctly harmful. As to its specific neutralizing power, this would have to be exerted very speedily, since the damage is so often done within 48 or 72 hours after the onset of symptoms."

Meltzer[125] has suggested the use of adrenalin intraspinally during the acute stage with the idea of lessening the congestion and thereby preventing to a certain extent the edema which occurs from the action of the poliomyelitis virus. Two cubic centimeters of a 1:1000 solution of adrenalin is injected every four to six hours after removing all the spinal fluid that comes away under pressure. However, its use is not recommended by the New York City Department of Health.

In cases with respiratory paralysis, however, it may have some value perhaps in giving a longer time for the immune serum to act.

In the acute stage the two important things are complete rest and the prevention of deformities. There is more harm done by trying to do too much than trying to do too little. During the period when there is any pain or tenderness there should be no attempt at massage, electricity or any attempts to force movements. Patients should have as complete rest and quiet as possible. There is no danger of the joints becoming fixed or ankylosis setting in and atrophy does not occur. By complete rest the cord will be given the best chance to begin repair. The prevention of deformities should be sought, especially "dropped foot." This condition can be prevented by supporting the bottom of the foot and raising the bed clothes. The posture in which the child lies in bed should be carefully watched. As deformities may begin during the first week, they can be largely prevented if every precaution is taken to keep the muscle at rest and in the normal position. After the tenderness and pain have disappeared it may be advisable to place the child in a warm salt bath. This can be done by lowering the child in a sheet into the tub. This gives opportunity for the easiest movement of the limbs without pain. No electricity nor massage should be used during the acute stage.

Treatment during the Convalescent Stage.—The convalescent phase may be said to begin when all the tenderness has disappeared. This usually takes several weeks, on the average perhaps from four to six weeks after the onset. The question arises at this time how long to keep the patient in bed. It must be remembered that it is not a good procedure to keep the child in bed too long. They are more likely to assume bad postures and to start the development of flexion deformities by being allowed to remain in bed for too long a period. Neither is this a good procedure from a psychologic standpoint. As has been pointed out by Lovett[126] it is wise to encourage the upright position, first because it "antagonizes evils of the permanent sitting position and also because the effort to balance on the feet instinctively excites to effort a large number of muscles not otherwise to be reached and is a valuable form of muscle training."

Whenever the patient can stand and walk without artificial assistance it is to be encouraged but where he is not able to walk, light braces should be used and crutches may be needed if the patient cannot walk without them. Deformities, however, should not be allowed to occur in standing or walking; stretching the muscles leads to deformities especially around the joints. In taking their first exercises or in first walking, it is important not to allow children to get fatigued. As Lovett[126] has pointed out "muscles are more often weakened than totally paralyzed." Therefore, it is very important to watch for over use or for fatigue. Therapeutic measures may be divided into massage, electricity, heat and muscle training.

1. Massage.—Massage is important during the early stages as it helps to keep muscle tone, stimulates the normal flow of blood and where there is stasis it empties out the blood from the veins. It probably does not increase the power of the muscle and it may do harm if it is not properly carried out or if it is done over too long a period so as to cause fatigue.

2. Electricity.—Many workers have advocated electrical treatment. The faradic current causes the muscles to contract. This may be a good way to stimulate muscle movement where one is unable to get voluntary movement but on the whole children do not take the faradic current well. Care should be taken that the current is not too strong. The positive pole should be put on the muscle to be stimulated and the negative pole some distance away or on the spine. The galvanic current has been used by some but it probably has little or no effect. However, it should be clearly borne in mind that by electricity very little as compared with muscle training can be achieved and often much good time is wasted by using electricity.

3. Heat.—Heat is of especial value where there is sluggish circulation and where the extremities tend to be cold and should be used before massage or exercises are undertaken in order to get the parts in as good local condition as possible before starting in with treatment at any given time. An electric pad is a very simple way of employing heat.

4. Muscle Training.—This is the most valuable measure in treatment. It should, however, be carried out by someone who is trained in this procedure. It is best to start with short periods and increase them as the muscle strength develops. Most gratifying results have been obtained by this method of treatment and probably far fewer cripples will be found from the last few epidemics due to the introduction of this method of treatment. As has been noted there are many more partial paralyses than complete and new routes for carrying impulses can be trained and brought under voluntary control. As has already been stated, with this treatment Lovett obtained the best results where it was carried out by an expert. Anyone having to handle a case is recommended to consult Lovett's book on "Treatment of Infantile Paralysis"[127] and Wright's book on "Muscle Training in the Treatment of Infantile Paralysis."[128]

At all times deformities should be prevented. It is better to prevent them than to correct them after they have occurred. This can only be accomplished by carefully watching cases during the convalescent and after period. The convalescent period or the period during which return of muscle activity may be expected will last for a period of from one to two years, though return to muscle activity has been noted after this period, as long as six or seven years and longer after paralysis.

The correction of deformities is a surgical orthopedic procedure and will not be dealt with here, so also the management of the chronic stage. During this stage operations of one type or another are indicated. Such operations should only be undertaken by those who understand statics and the function of muscles, in other words, by orthopedic surgeons. The application of proper braces and operations can only be successfully carried out by those who are familiar with this type of treatment.

Poliomyelitis from the Public Health Standpoint.—Although our knowledge to date of the means of transmission, period of incubation and length of contagion are not clearly defined, still much has been gained of importance in the management of epidemics in this country. The New York Epidemic of 1916 was carefully studied from this point of view and certain fairly definite conclusions have been arrived at.

1. "That hospitalization when prompt and applied to all but moribund cases probably checks the spread of the disease and determines a lower incidence under similar housing conditions than where removal to hospitals is delayed and many cases are left in their homes." In support of this the official reports of the Department of Health point out that in Brooklyn only 46 per cent. of the cases which were suitable for removal to hospitals were taken from their homes to isolation hospitals, while in the Borough of Manhattan 96 per cent. were hospitalized and though they do not claim that all other factors have been excluded yet it is certainly suggestive that in Manhattan, where there is a greater population per acre, the case incidence of the disease was 0.94 per 1000 of population, while in the Borough of Brooklyn it was 2.24 per 1000 of population.

2. "Isolation of groups of children from contact with other children or adults, even when carried out in the midst of areas where the disease is prevalent, suffices to protect such children almost absolutely from infection, in spite of the use of identical water and food supplies and exposure to the same atmospheric conditions and winged insects." This has already been quoted as showing the bearing these factors had on the condition (Barren Island incident).

In relation to the control of the disease the New York report says: "In preparing to meet and control the disease, if or when it reappears, we must, until new information as to carriers and means of transmission is forthcoming, rely upon early diagnosis, prompt notification, hospitalization or equivalent home isolation, a well informed public and an alert profession."

The following is the general scheme for meeting the situation as outlined by the New York City Department of Health, July, 1916:

"Information for the Public.—Infantile paralysis (poliomyelitis) is a catching disease. How it is spread is not yet definitely known. In most cases the disease is probably taken directly from a sick person, but it may be spread indirectly through a third person who has been taking care of the patient or through children who have been living in the same household.

The early symptoms are usually fever, weakness, fretfulness or irritability, and vomiting. There may or may not be acute pain at this time. Later there is pain in the neck, back, arms or legs, with great weakness. If paralysis is to occur it usually appears from the second to the fifth day after the sickness begins. Many cases do not go on to paralysis.

The germ of the disease is present in discharges from the nose, throat and bowels of those ill with infantile paralysis even in the cases that do not go on to paralysis. It may also be present in the nose and throat of healthy children from the same family. Do not let your children play with children who have just been sick or who have or recently have had colds, summer complaint, etc. For this reason children from a family in which there is a case of infantile paralysis are forbidden to leave their home. If you hear of their doing so report it at once to the Department of Health.

"**How to Guard against the Disease.**—In order to prevent the occurrence of this disease, parents should observe the following rules:

Keep your house or apartment absolutely clean.

Go over all woodwork daily with a damp cloth.

Sweep floors only after they have been sprinkled with sawdust, old tea leaves, or bits of newspaper which have been thoroughly dampened. Never allow dry sweeping.

Screen your windows against flies, and kill all flies in the house.

Do not allow garbage to accumulate, and keep pail closely covered.

Do not allow refuse of any kind to remain in your rooms.

Kill all forms of vermin, such as bed-bugs, roaches, and body lice.

Pay special attention to bodily cleanliness. Give the children a bath every day and see that all clothing which comes into contact with the skin is clean.

Keep your children by themselves as much as possible. Do not allow them to visit moving picture shows or other places where children may gather.

Children should not be kept in the house; they should be out-of-doors as much as possible, but not in active contact with other children of the neighborhood. Do not take them on a street car, unless absolutely necessary, or shopping.

"Do not allow your children to be kissed.

It is perfectly safe to let your children go to the parks and play grounds if only two or three of them play together; they should not play in large groups, and you should not let them come into contact with children from other parts of the city.

Remember that children need fresh air in the summer time, and outdoor life is one of the best ways to avoid disease.

If there is a public shower bath in a school in your vicinity, send the older children there every day for a shower bath. This is perfectly safe and will help keep them in good health.

Give your children plain, wholesome food, including plenty of milk and vegetables.

Keep the milk clean, covered and cold. Do not allow the milk or any other food to be exposed where flies may alight upon it.

Wash well all food that is to be eaten raw.

In Case of Sickness.—Remember that during the hot weather children are apt to have stomach and bowel troubles. If your child is taken sick with loose movements of the bowels, or with vomiting, do not at once fear that it must be infantile paralysis; it may be simply digestive disturbance. Give the child a tablespoonful of castor oil and plenty of cool water to drink, and *send for the doctor at once.*

If you cannot afford a doctor's services, telephone the Department of Health and one will be sent free of charge.

If a doctor or nurse from the Department of Health visits your home, give them all the information you can. They are sent to show you how to keep your children well.

Do not give your children patent medicines or buy charms of any kind to ward off disease. *The best preventive is cleanliness and strict observance of the rules that have been given.*

"**Information for Physicians.**—*Early Diagnosis.*—The attention of physicians is called to the necessity of an early diagnosis of all cases of poliomyelitis. Early recognition and strict quarantine are the chief weapons against the disease.

Reporting of Cases.—All suspicious cases must be at once reported to the Department of Health by telephone, to be followed within 24 hours by a written report. The ability of the Department of Health to limit the spread of the infection depends upon the immediate reporting of every suspicious case.

Age of Persons Affected.—It should be remembered that this disease may occur at all ages, although the great majority of the cases are found in children between the ages of one and five years.

Type of Disease.—Peabody, Draper and Dochez, of the Rockefeller Institute, give the following classification of the disease:

1. The non-paralytic or so-called abortive cases.
2. The cerebral group, with spastic paralysis.
3. The bulbospinal group.

Methods of Infection.—The experiments of Landsteiner and Popper in Germany; Kling, Pettersson and Wernstedt in Sweden, and of Flexner and Noguchi in this country have proved that the disease is transmitted by the secretions of the nose and mouth and the bowel discharges of an infected person. The infection is transmitted through the mouth, tonsils and nasal mucous membrane.

Contacts and Carriers.—It must be remembered that while the transmission of the *disease* from a patient to other members of the same family is not usual, transmission of the *virus* is common. Experience warrants the assumption that in addition to direct contact the disease is spread by carriers, usually children, who are themselves immune but who harbor the infective material in their nasal or mouth secretions.

Symptoms.—Early symptoms to be regarded as suspicious are: Fever, vomiting, slight diarrhea, listlessness, unusual fretfulness, and drowsiness. Later and more characteristic symptoms are: The appearance of weakness in any extremity, skin and muscular sensitiveness, spinal pain, especially on flexion, apparent or real rigidity of the neck muscles, Kernig's and MacEwen's signs.

Course and Duration of Disease.—Paralysis appears usually before the sixth day of the illness; it may occur as early as the first day. Other symptoms, especially spinal and muscular pain and rigidity and skin sensitiveness, rarely persist.

Non-paralytic or So-called Abortive Cases.—Non-paralytic or so-called abortive cases are very frequent. In some epidemics they constitute from 25 to 50 per cent. of the diagnosed cases. The children have the early symptoms just mentioned, perhaps also the muscular tenderness and spinal pain. If carefully observed it may be noticed that they develop a paralysis of one or more groups of muscles, but that instead of the paralysis continuing it all disappears within a few hours. It is obvious that the recognition of such cases is of extreme importance in controlling the spread of the disease. The diagnosis of such cases is greatly facilitated by an examination of cerebrospinal fluid obtained through lumbar puncture.

General Care of Patient.—Complete rest is of the utmost importance for either paralyzed or weak muscles for the first five or six weeks. Every effort must be taken to make this rest complete. The limb must not be allowed to drag on a paralyzed muscle. It should be supported by pillows or pads or bandages. There seems to be a greater tendency to atrophy if casts are used. A dropped foot may be supported by a sandbag or pillow; small rolls placed under the knee often hold the leg in a more comfortable position. The weight of the clothing should be kept off the legs by hoops or other device. If the head is somewhat retracted and the patient desires to lie on his back, he may sometimes be made more comfortable by a small pillow placed under the shoulders, allowing the head to fall back. The value of electricity for treatment in the first six weeks is very doubtful. In many instances it may do harm. Massage or passive movements should not be begun for at least five or six weeks and then should be used with great care. In cases that show a tendency to clear up rapidly the child should be kept in bed for some time after the ability to use the muscles returns. It should never be encouraged to try to stand or to use the muscles otherwise until a considerable time has passed.

Period of Incubation and Duration of Disease.—The incubation period has been officially set at two weeks. Non-immune, infected persons usually manifest symptoms of the disease in from 5 to 10 days after exposure. The average period of incubation is seven days. The early symptoms, noted above, usually last from one to seven days. Quarantine of the patient will be maintained for a period of at least eight weeks.

Prevention of Spread of Infection.—1. The children from an infected family will be confined to the house.

2. During the continuance of an epidemic of poliomyelitis children should not be allowed to congregate in public places.

3. Fresh-air outings or vacation camps are allowed, if kept under competent medical supervision, with an adequate physical examination of each child before enrollment and the exclusion of any child from an infected family.

4. Absolute cleanliness of all homes is essential; such cleanliness should include:

(*a*) Screens in all windows.

(*b*) Flies kept out of all rooms.

(*c*) Thorough cleanliness of all floors, woodwork, bedding, and clothing.

(*d*) Avoidance of dust (all sweeping should be done after the floors have been sprinkled with sawdust, bits of paper, or tea leaves, all thoroughly moistened).

(*e*) Garbage cans kept closely covered and washed out in hot soapsuds after they have been emptied.

(*f*) No refuse, either of food or other waste, allowed to accumulate.

5. Personal habits of cleanliness are essential; the hands should be washed before each meal, after each visit to the toilet, and before going to bed. Children should be warned about putting the fingers into the mouth or nostrils.

6. When sneezing or coughing, a handkerchief should be held over the mouth. Kissing of children is also a dangerous practice and should be avoided.

Procedure to Be Followed in Each Case.—1. Isolation of patient: Complete isolation of the patient must be maintained until terminated by order of the Department of Health.

2. A separate room must be provided for the patient. No one must be allowed in this room except the attending physician, the nurse, and the representative of the Department of Health.

3. Care of patient's room and surroundings: (*a*) All rugs, carpets, draperies, and unnecessary furniture must be removed before the patient is placed in the room.

(*b*) All windows must be screened.

(*c*) The sick room must be kept well aired at all times.

(*d*) The woodwork must be wiped daily with damp cloths. Under no circumstances must the floor be swept when it is dry. It should be sprinkled with sawdust, bits of newspaper or tea leaves, all thoroughly moistened and then carefully swept so that no dust may arise.

(*e*) Toys and books used by the patient must be destroyed by burning after recovery or death.

(*f*) Household pets must not be allowed in the room.

4. Care of bedding: All cloths, bed-linen, and personal clothing which have come into contact in any way with the patient must immediately be immersed in a 5 per cent. solution of carbolic acid and allowed to soak for three hours. They may then be removed from the room and must be boiled in water or soapsuds for 15 minutes.

5. Care of discharges from body: A sufficient supply of gauze or clean linen or cotton cloth must be provided and all discharges from

the nose and mouth of the patient received on these cloths. After use they must be immediately burned or boiled. Bowel discharges and urine must be covered at once with chlorid of lime and then disposed of by emptying into a water-closet.

6. Care of utensils used by patient: Plates, cups, glasses, knives, forks, spoons and other utensils used by the patient must be kept for exclusive use and under no circumstances removed from the room or mixed with similar utensils used by others. They must be washed in the room in hot soapsuds and then rinsed in boiling water. After use the soapsuds and water must be thrown into the water-closet.

7. Nurse: A trained nurse or competent attendant must be in sole attendance upon the patient. She must not be allowed to mingle with the rest of the family but must be isolated with the patient. The hands of the nurse must be carefully washed in hot soapsuds after each contact with the patient and before eating.

8. Termination of case: After the case has been ordered terminated by the Department of Health the following procedure must be followed:

(*a*) The entire body of the patient must be bathed and the hair washed with hot soapsuds. The patient should then be dressed in clean clothes (which have not been in the sick room during the illness) and removed from the room.

(*b*) The nurse should also take a bath, wash her hair, and put on clean clothes before mingling with the family or other people.

Action Taken by the Department of Health in Each Case.—Placarding: Every house will be placarded without exception. In private houses one placard is placed on the street front (outside of house), and one placard is placed on the door entering the room patient is in. In tenements three placards are affixed, one on street door, one in entrance hall, and one on door of apartment. All placards are dated.

Quarantine: In all families where a case of poliomyelitis has occurred all other children under 16 years except those who have had the disease are to be quarantined in the home until two weeks after the termination of the case by death, removal, or recovery.

The patient, whether at home or in hospital, must be quarantined for eight weeks from onset of disease.

Children under 16 years of age who have been, but no longer are, exposed to infection will be quarantined for 14 days.

Removal to hospital: No case is to be left at home unless the following conditions are complied with:

(*a*) There must be a private physician in attendance regularly.

(*b*) Persons attending patient must obey quarantine rules; must not do any housework, marketing or leave premises.

(*c*) Patient and attendant must have separate rooms.

(*d*) All windows of rooms used by patient must be screened.

(*e*) The family must have a separate toilet for its exclusive use.

(*f*) Quarantine regulations must be strictly observed by patient and other children.

Deaths: In case of death prompt burial is required, the coffin must be sealed as in deaths from other contagious diseases, and the funeral must be strictly private. Church funerals are prohibited.

Spinal puncture: Physicians desiring the services of a consultant to perform lumbar puncture and report on the examination of spinal fluid should telephone to the Department of Health."

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CHAPTER CLXXXI

MULTIPLE NEURITIS. THE NEURALGIAS. HERPES ZOSTER (SHINGLES). GROWING PAINS. SPASMS. POLYMYOSITIS. PROGRESSIVE MYOSITIS OSSIFICANS. ISCHEMIC MUSCULAR PARALYSIS AND MUSCLE CONTRACTURE. TUMORS OF THE NERVES

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MULTIPLE NEURITIS

Multiple neuritis or polyneuritis is a general disseminated inflammation or degeneration of the nerves, symmetrical in distribution, generally affecting the nerves of the limbs in their terminal branches.

Cases may be classified etiologically into:

1. Toxic cases due to the action of an exogenous toxin. Such toxins are alcohol, carbon monoxid gas, bisulphide of carbon, nitrobenzol, the coal tar products and arsenic, lead, mercury, copper, phosphorus and silver.

2. Infectious cases due to some agent acquired or developed within the body as a result of diphtheria, typhoid fever, grippe, scarlet fever, measles, whooping-cough, smallpox, and the epidemic form of beriberi.

3. Cases due to general diseased states of the body as rheumatism, marasmus, general malnutrition, consequent upon tuberculosis or lues and, finally,

4. Cases due to exposure to cold or developing spontaneously without known cause.

Multiple neuritis is essentially a disease of early adult life. It is rare in childhood if we except the cases following diphtheria. Starr[1] in 154 cases of multiple neuritis found none under 15 years of age. In 94 cases of Remak,[2] nine were between 10 and 20 years of age. Bruns,[3] in a practice extending over 25 years, saw but two cases in children. Thomas and Greenbaum,[4] reviewing the literature in 1907, tabulated 138 cases of non-diphtheritic multiple neuritis in children and reported eight cases of their own. Of the 138 cases tabulated, three followed grippe, eight measles, five typhoid fever, seven whooping-cough, one chicken-pox, six scarlet fever, and 15 were miscellaneous, making a total of 48 cases following infections. Of the toxic cases, 19 followed lead, five alcohol, one carbon-monoxid, 19 arsenic (chorea cases), four arsenic (non-chorea cases), making a total of 48 cases. There were 38 cases of unknown origin and also included in the series were nine cases of ascending palsy.

Of the eight cases reported by them, three followed typhoid fever; the other five were of unknown etiology.

Cases following typhoid fever have been observed by Bruns, Jones,[5] and Humphreys;[6] following mumps, Remak and Joffroy;[7] whooping-cough, Möbius,[8] Surmay,[9] Mackey,[10] Gunion,[11] Aldrich,[12] and Eshner;[13] influenza, Bruns, Schwarz; bronchopneumonia, Owen;[14] scarlet fever, Seifert,[15] Remak, and Basette;[16] varicella, Donet; variola, Leon;[17] and measles, Denarié.[18]

A number of cases have been attributed to malaria by Gibney,[19] Sachs,[20] and Browning.[21] Dana[22] considers malarial multiple neuritis as probably being a form of beriberi. In the case of Gibney the pathology was found to be a congestion and edema of the cord along with other changes in the peripheral nerves.

Mention should be made of acute pernicious multiple neuritis which comes on suddenly, progresses rapidly and causes death in a few days or weeks. These cases usually show the ordinary symptoms of neuritic paralysis and probably are due to the same cause as poliomyelitis, occurring along with this disease during epidemics. Medin[23] saw six cases in 1887 and two in 1895. Hammond[24] reported 10 cases in an epidemic in 1895, occurring in children from four and one-half months to four and one-half years of age. Caverly[25] and MacPhail,[26] in a report of 126 cases of infantile paralysis, note that in some there were elements strongly suggestive of multiple neuritis, and Blackall[27] in 1904 reported an epidemic of six cases.

Recurrent multiple neuritis has been observed by Rabinowitz.[28]

Special emphasis must be placed upon the fact that a considerable proportion of the cases of multiple neuritis occurring in children follow no known cause.

Whatever may be the cause of multiple neuritis, the symptoms, with some modifications, are the same in all cases. The disease often begins with prodromata sometimes lasting weeks. The patient complains of numbness, slight fever, tingling in the extremities and sensations of heat and cold; then there follows a weakness which may occur rather suddenly or subacutely. At times it is associated with febrile disturbances; rarely with convulsions. The pains and weakness increase; the muscles and nerves become very tender, while at the same time the skin may be reddened or slightly edematous. In a day or two the patient is unable to stand. In a week or two there may be a complete paralysis in the anterior tibial group or extensors of the wrists.

An acute febrile polyneuritis has been described by Osler which could be attributed to no known cause. On the other hand, many cases are unassociated with febrile disturbances.

The paralysis is a flaccid one. It is followed by atrophy and associated with trophic changes. There is an alteration of electrical conditions, varying from an early loss of faradic and diminution of galvanic stimulation, to complete reaction of degeneration. The distribution of the paralysis is usually symmetrical. Certain groups of muscles are more readily affected than others, as the extensors of the foot and wrist are frequently involved in the early stages of the disease.

One of the most characteristic features of multiple neuritis is the association of sensory changes with paralysis of the lower motor neuron type. There is some tactile anesthesia, often with hyperalgesia. Later, the temperature and pain sense are lessened and slowed in transmission. The anesthesia is either patchy, or may occur diffusely. Muscle and articular sense are frequently lost, especially in the pseudotabetic or ataxic forms of the disease. The deep reflexes are usually diminished or absent, and the superficial reflexes are frequently lost.

The more distal parts of the extremities are, as a rule, the first to be affected; in rare cases the motor cranial nerves evidence the first signs of the disease. The facial, oculomotor, abducens, patheticus, trifacial, auditory, glossopharyngeal, vagus, and hypoglossal nerves have all been involved in various cases.

The bladder may occasionally be affected for a short time; the other visceral centers not at all.

The course of the disease varies, depending on the intensity of the cause. As a rule, the symptoms increase during the first four to six weeks, and then diminish. While the disease is progressing, the sensory symptoms are particularly prominent. The motor paralysis disappears much less quickly than the disturbances of sensation. After a time if the paralysis is great, contractures occur; the feet are extended, the waist and legs are flexed on the thighs and may be almost immovable. The deformities arising from this disease are quite as serious and as persistent as those in poliomyelitis.

Pathologic Anatomy.—Inflammatory and degenerative changes in the peripheral nerves form the pathologic basis of the disease. The neuritis develops chiefly in the peripheral ramifications of the nerves. Inflammation is less marked than degeneration and atrophy. Swelling and redness of the nerves and hyperemia of their sheaths are present macroscopically. Perineuritic and interstitial changes are either slight in extent or are generally absent. Pathologic affections vary from slight peri-axillary neuritis to complete degeneration of the nerves. Evidence of regeneration occurs along with the degenerative changes. The myelin frequently is disintegrated into clots and granules. In some cases the muscular tissue is slightly involved; the fibers are smaller and paler; and the transverse striations are less distinct. The nuclei of the fibers and interstitial tissues likewise show changes.

Diagnosis.—The diagnosis of multiple neuritis is usually an easy matter. When anesthesia and ataxia are the most prominent clinical features, the symptoms resemble those of juvenile tabes dorsalis. The acute onset, the absence of Argyll Robertson pupils and bladder disturbances, some evidence, however slight, of involvement of the motor system, and the serologic examinations tend to differentiate this condition clearly.

It is differentiated from Friedreich's ataxia by the presence of deformities, scoliosis, pes cavus, the disturbance of speech, the hereditary character, the mental changes, and the gradual onset.

It is differentiated from anterior poliomyelitis by the absence of intense tenderness of nerve trunks to pressure, by the asymmetrical distribution of paralysis, and by the absence of sensory findings. (Of course lead paralysis occupies a unique position in this respect, in that it usually is purely motor, and in this regard, therefore, does not differ from poliomyelitis.) Involvement of the cranial nerves is rare in poliomyelitis. The recurrence of increasing motor disability is evidence of peripheral neuritis. As a further distinction between the two diseases, it may be pointed out that in poliomyelitis diminution in galvanic irritability comes on only after weeks or months. Although spinal fluid changes have been reported in multiple neuritis, pleocytosis and increase of globulin is more constant in poliomyelitis.

Trichinosis may give rise to symptoms similar to multiple neuritis. Its diagnosis can usually be made from the early gastro-intestinal symptoms, fever, muscular pains, followed by the characteristic muscular swelling, difficulty in moving the legs, which are involved by edema. Great increase of eosinophilic polymorphonuclear leukocytes in the blood is suggestive of trichinosis. Finally, the discovery of the organisms in the muscles and spinal fluid makes the diagnosis absolute.

Mention may be made of the remote similarity between multiple neuritis and progressive muscular atrophy, the dystrophies, myelitis, Pott's disease, periarteritis nodosa, rickets, and hysteria.

The **prognosis** is good. In Thomas and Greenbaum's series, 58 made a complete recovery, 46 a partial recovery; in 33 cases the outcome was not mentioned; 14 cases died. Of these last the neuritis in two cases was due to alcohol; in one to lead, one to angina follicularis, one to syphilis, six to ascending paralysis—which probably should not be included in the multiple neuritides—and one to an unknown cause.

Treatment.—In the treatment of the multiple neuritides, the first indication is, if possible, to remove the cause. Alcohol, of course, should be withdrawn. In lead poisoning further contact with the offending toxin should be prevented. Further elimination of the lead may be promoted by the use of iodid of potassium, by the administrations of aperients, and by baths—preferably sulphur.

For cases in which the source of poisoning is endogenous, Oppenheim strongly recommends diaphoresis. If the patient's strength permits, the secretion of sweat may be stimulated by hot baths; otherwise, by hot packs, hot air applications along with hot drinks and aspirin. In all forms of neuritis a strengthening diet is indicated.

If the pain is not relieved by warm fomentations, antineuralgic drugs may be used. Great reluctance should be exhibited and care exercised in the prescription of opium or its derivatives. In treating children this drug should be avoided entirely. General tonic medication may be prescribed, cod-liver oil, iron, quinin, and strychnia.

During the first stages of the disease absolute rest in bed is imperative. Steps must be employed to prevent the stretching of any of the paretic muscles; light splints may be applied to keep the extremities in a neutral position. At the beginning, active and even passive movements must be avoided as much as possible. When the neuritis has reached its height and the condition has either become stationary or shows its first sign of improvement, mechanical and electrical treatment becomes advisable. Labile galvanic stimulation should be used, employing the positive pole for the active electrode and a current no stronger than is necessary for producing a muscular contraction. As the reaction to faradism returns, labile faradic stimulation may likewise be employed. Massage is of great value but must be used with care so as to avoid bruising the tender muscles. Passive movements and gradually increasing resistive exercises may be employed later in the course of the disease, always avoiding all undue fatigue and violence. Subcutaneous injections of strychnin several times a day are of service in persistent paralysis.

Measures should be taken to prevent contractures. If, unfortunately, they do occur, they must be treated according to well known orthopedic procedures. The uncontrolled and negligent use of braces is particularly to be warned against as long lasting muscular disuse is not conducive to recovery of function.

Residual paralysis may be treated by various surgical procedures, as tendon, muscle, or nerve transplantation.

Diphtheritic Multiple Neuritis.—Diphtheritic multiple neuritis or paralysis manifests itself in several forms. Those cases in which we observe signs of paralysis of the soft palate, of the muscles of deglutition, of ocular accommodation, and where perhaps there are also signs of cardiac involvement, have been grouped as localized or circumscribed forms of diphtheritic paralysis. They are differentiated from those cases in which a generalized affection of the nerves has occurred and which are classified as the polyneuritic or general types of paralysis. There is, however, no sharp line of demarcation between these two groups, and the extension of the circumscribed forms into the generalized, pathologic, and neuritic phenomena is frequently indicated by such objective signs as changes in the deep reflexes of the lower extremities. In addition, it should be noted that polyneuritic forms frequently develop after the appearance and course of a localized paralysis.

To the first form belong the isolated palsies of the palate and the muscles of ocular accommodation. The polyneuritic forms may assume the character of an amyotrophic type or, through special selection of the sensory fibers, conduction and deep sensation, give rise to an ataxic form recalling the picture of pseudo-tabes.

Frequently the condition is characterized by involvement of the vagus and phrenic nerves accompanied by disturbances of respiration. In certain severe cases all the above disturbances may be present at the same time and in addition to involvement of the external ocular muscles, there may also be disturbances of the facial, trigeminal, or hypoglossal nerve. Very often these cases begin first as an isolated palatal palsy from which recovery takes place followed at some later time by the other nervous disturbances.

Postdiphtheritic paralysis occurs in from 9 to 12 per cent. of the cases of diphtheria.

In 2448 cases collected by Sanne,[29] paralysis was noted in 11 per cent. Lennox Browne[30] in a series of 1000 cases reported 14 per cent. In 1071 cases belonging to pre-antitoxin days, Goodall[31] found paralysis in 11.7 per cent. of the whole number after deducting a mortality of 33.8 per cent. In 3384 cases treated by antitoxin, which were collected by the American Pediatric Society,[32] paralysis was observed in 9.7 per cent. The figures in the report of the Metropolitan Asylums Board of London, based upon a collection of 98,238 cases, are rather high—18.5 per cent. (Northrop[33]). On the other hand, Henoch[34] gave as low a figure as 5 per cent. Recently, Rolleston[35] states that of 2300 cases of diphtheria which he observed, 20.7 per cent. showed some type of paralysis. He included in his calculation those cases showing the slightest evidence of neuritic change. The fact that the paralysis in his cases never lasted over nine days indicates the mildness of the types he describes.

Postdiphtheritic paralysis attacks adults as frequently as children. According to Sachs,[36] it is relatively less frequent in children. Rolleston, however, basing his data upon much better ground, says that it is commoner in children. In 267 cases of diphtheria, out of a total of 1350, the knee-jerks were absent. In 196, the Achilles jerk was lost. Of this number only four were over 14 years of age, whereas 149 cases were above that age. In a series of 1316 cases at the Park Hospital Meyers[37] reported 275 cases with 80 deaths. Of these, 104 occurred in children under five; 138 from five to eight; and 33 in children above 10. The males predominated. The average day of the appearance of paralysis was the twelfth; the earliest the fourth. In Rolleston's series of 1500 cases, published in 1909,[38] paralysis rarely occurred before the third week, except from severe diphtheria in which palatal and cardiac paralysis occurred. The usual time of cycloplegia was four to five weeks. Palsies of the lips, face, neck muscles, extremities, as well as vesical and rectal troubles, rarely occurred before the fifth or sixth week.

Treatment by means of antitoxin has resulted in changes in the percentage of cases of paralysis, because many cases which would formerly have ended fatally now go on to convalescence, and which as a result of the severity of the infection, develop a paralysis. In 1898 the London Clinical Society reported that 22 per cent. of a series of 6633 treated cases resulted in paralysis, whereas in untreated cases there was an incidence of 11 per cent. Starr, like Sachs and others, thinks that paralysis is not as frequent a sequel in cases treated by antitoxin as in those cases untreated by this method. Rosenow and Anderson[39] have shown experimentally that one unit of antitoxin injected on the first day prevents paralysis in guinea pigs, whereas 4000 units do not suffice 48 hours after diphtheritic infection. Ransom[40] is led to believe that if given early, antitoxin influences favorably subsequent paralysis.

Opinion differs as to the correlation between the severity of the initial attack and the subsequent paralysis. It is true that the paralysis bears no absolute relation to the severity of the diphtheritic infection. Typical cases are seen which set in after an infection so mild that it may have been considered a slight cold or else overlooked entirely. It has been demonstrated, however, both experimentally and clinically, that in general a definite relation exists between the severity of the primary infection and the subsequent paralysis. Ransom has shown that the severity of the paralysis is proportionate to the amount of toxin administered. Rolleston, in a series of 100 cases, points out the fact that the frequency and degree to which the Achilles jerks are involved have a direct connection with the character of the initial attack, corresponding in this regard to the paralysis and albuminuria. In his recent investigation he is able to corroborate this conclusion. Woolacot[41] collected 829 consecutive cases of uncomplicated diphtheria, taken from the report of the Metropolitan Asylums Board for 1898, and divided them into three classes—severe, moderate, and mild. His results show that paralysis occurs most frequently in the severer cases, thus substantiating Ransom's experimental findings.

The first and most common symptom of paralysis following diphtheria is palatal palsy. In 171 cases of diphtheritic paralysis observed by Ross,[42] the palate was affected in 128; the eyes in 77 (the muscles of accommodation were here affected); the lower extremities in 113; the upper extremities in 60; the trunk and neck in 58; and the muscles of respiration in 33. Of 328 cases reported to the American Pediatric Society, the distribution was specified in 187. Of this number 120 involved the throat, palate, pharynx, larynx; in 14 the extremities; in 11 the eyes; in 32 the heart; and in eight cases the paralysis was general. In the series published by Goodall, the palate alone was first affected in 66 per cent. of the 125 cases, and in combination with other muscles it was involved in 12 per cent. more. The affection of the throat is, therefore, the most common. It may occur alone or be followed by paralysis of other parts—the eyes, the extremities, the trunk, or the neck. In most of the throat cases, the uvula and the soft palate are involved.

These cases may occur as an early paralysis (diphtheritische Frühlämung) of Baginsky.[43] In the severest form of diphtheria, the paralyses affect the velum palati; the speech becomes nasal and swallowing difficult, so that ingested fluids are regurgitated through the nose. Food may pass into the trachea and cause death by pneumonia. The paralysis is usually bilateral, although it may be unilateral. It has an evil significance in its association with cardiac paralysis.

On the other hand, it may occur as a late paralysis, coming on 14 days to three or five weeks after the primary infection. The symptoms in both instances are similar. The soft palate becomes perfectly flaccid and droops, and there is an absence of the reflex contraction, which accounts for the nasal tones as well as for the regurgitation of fluids through the nose. In many cases there is paresis of the pharynx and esophagus as well as of the soft palate. Dysphagia may result, which in connection with hypesthesia of the entrance to the larynx, may cause food to find its way into the trachea. Severe coughing frequently ensues, and aspiration pneumonia is a common sequel.

Hoarseness and aphonia due to paralysis of the vocal cords frequently occur. Hypesthesia of the palate and disappearance of the palatal reflex are an accompaniment of the palatal paralysis. The paralysis is atrophic, and reaction of degeneration may be present.

Paralysis of the accommodation of the eyes is of relatively infrequent occurrence and appears later in the course following the primary infection. This affection makes it almost impossible for the patient to read fine print or to sew, although he may see well as to distance. Contraction during accommodation may be observed, although the act of accommodation itself is carried out imperfectly. The ciliary muscle is often paralyzed, and the reaction of the pupils to light is sluggish. The pupils may be unequal.

Much less frequently does the paralysis involve the external muscles. In such an event, however, the sixth cranial nerve is the most likely to be affected, and the fourth, the least. Complete oculomotor paralysis is rare. Ptosis and paralysis of one or more muscles supplied by the third nerve have frequently been noted.

Intermediate between the purely localized forms and the general polyneuritic types are the cases already referred to, in which the pharynx and larynx may be involved in addition to the palate. Changes in the deep reflexes are an early manifestation of general neuritic involvement. The absence or diminution of the knee-jerks had been noted by many. Rothe[44] found among 51 cases, 15 in whom the knee-jerk was arrested. Remak, among 78 cases in which there was paralysis of the soft palate, found 40 in whom the knee-jerk was absent. In the majority of cases the disappearance of the deep reflexes follows the paralysis of the soft palate, but Baginsky reports a disappearance of the knee-jerk on one or both sides as the first sign of diphtheritic paralysis. Rolleston[45] in 100 cases of diphtheria, examined from 20 to 63 days following the onset, found the Achilles jerk affected in 47 cases, absent in 20, and sluggish in 27. The knee-jerk was lost in 31 and sluggish in 38.

According to Monti,[46] Bristowe,[47] and other authorities, exaggeration of the patellar reflex and even ankle clonus may appear before the signs of paralysis, but such an occurrence is inconstant. Rolleston reports a Babinski phenomenon in 19.6 per cent. of acute cases. If the heart fibers of the vagus be involved a bradycardia appears. It is undoubtedly true that many disturbances of the heart are due to an involvement of the myocardium itself. A weak, irregular, intermittent, or rapid action may be observed. The first sound may be faint, short, associated with a systolic murmur. When there occurs, along with cardiac symptoms, an anesthesia of the palate and pharynx, an absence of palatal and pharyngeal reflexes, and paresis of the vocal cords, it is probable that the vagus itself is involved.

Involvement of the nerves of the cervical cord is a very common occurrence. The head can be balanced only on the neck, so that when the body is turned from side to side, the head falls, the muscles being unable to support it against the force of gravity.

The irregularities of respiration asssociated with heart symptoms are characteristic of vagus affections. According to Meyers, diaphragmatic paralysis occurred in 7.6 per cent. of 1316 cases. The symptoms were depression of the abdomen during inspiration and bulging during expiration. Respiration was rapid and panting, and asphyxia occurred on the slightest provocation.

When, as above outlined, these various symptoms are present in a single case, the child becomes a helpless, pitiful spectacle. When the muscles of deglutition are involved, the saliva either collects in the mouth or dribbles. This weakness occurs especially when there is facial paralysis or when swallowing is attempted in the presence of palatal palsy or anesthesia. The fluid, when it finds its way into the larynx, gives rise either to coughing, or when there is paralysis of the muscles of respiration, to a severe dyspnea. Such patients, because of inanition or aspiration pneumonia, have a rapid and usually fatal course. Attention has been called to a specially rapid and fatal form of paralysis involving the respiratory muscles which is accompanied by Cheyne-Stokes respiration and edema of the lungs (Baginsky and Bruns).

Two forms of the polyneuritic type may be found: the one regarded by Oppenheim[48] as the most frequent is the amyotrophic, while Bruns and Remak, on the other hand, have had the most experience with the ataxic or psuedotabetic type. Amyotrophic neuritis has its onset in five to seven weeks after the initial infection. It frequently follows a palatal or accommodation paralysis, though at times it may accompany these disturbances. It is ushered in by numbness, tingling, formication, seldom by pain; then followed by weakness and paralysis of the musculature. The paralysis is usually symmetrical and attacks by preference the lower extremities, either the peroneal muscles alone, or the peroneal and tibial groups of muscles together. There may appear more generalized forms in which the upper and lower extremities as well as the trunk are involved. In the upper extremities, the paralysis usually picks out the shoulder muscles and the interosseii of the fingers. There is a notable absence of any predilection for the extensors of the wrists. Very frequently an involvement of the muscles of the trunk gives to these children, when attempting to arise, the appearance of a pseudohypertrophic dystrophy. If the patient is able to walk, a steppage gait is apparent. The paralysis is degenerative in character, with atrophy, flaccidity of the muscles, and quantitative changes in electrical irritability. The latter, however, are not pronounced in character and seldom show complete reaction of degeneration. Incomplete reaction of degeneration and changes in faradic reactions are most common. It must be remembered, however, that in these children a complete electrical examination is frequently impossible.

Sensory disturbances are shown objectively, less by temperature and sense of pain than by contact, particularly by sense of position and other deep sensations. There is a marked decrease of vibratory sense. A delay of pain sense has rarely been observed. Vasomotor, trophic, and secretory changes are rare, although Oppenheim speaks of edema. The functions of the bladder and rectum are usually undisturbed. In extremely severe cases, facial paralysis may occur, either as a mono- or diplegia facialis, and sensory and motor trigeminal paralysis may ensue, as in the patient whose illustration is here presented

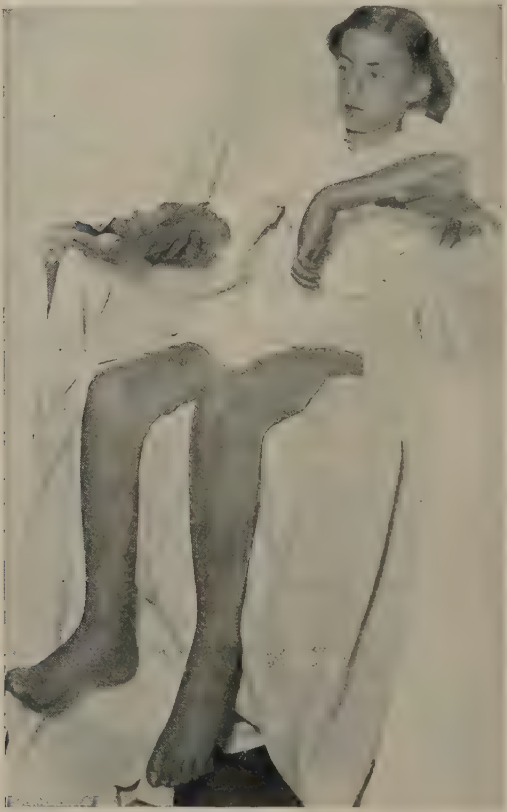

Fig. 62.—Illustration of a case of multiple neuritis or paralysis following diphtheria, showing paresis of the right side and paralysis of the left with a left facial palsy and involvement of the right trigeminus which occurred upon a late palatal palsy. (This case was seen through the kindness of Dr. Davies and Dr. Doepp of Blue Island, Illinois.)

(Figs. 62, 63). Even hypoglossal paralysis may occur; while auditory neuritis and optic neuritis have been noted by some observers.

The ataxic or pseudotabetic form develops in a like manner to the amyotrophic. The pure type of this form with absence of muscular weakness, atrophy, and reaction of degeneration is very rare, although it has been observed (Remak, Bruns). In these forms very marked symptoms of ataxia are found. The gait is uncertain and stamping. The movements lack coördination, especially when the eyes are closed. Romberg's sign is present. Uncertainty of movement is likewise demonstrable in the upper extremities and may be elicited by the finger to nose test and by the impossibility of employing the ordinary finer movements employed in writing, buttoning the clothes, etc. Objective sensory disturbances may be missing. Disturbance of deep sensibility, on the other hand, is rarely absent.

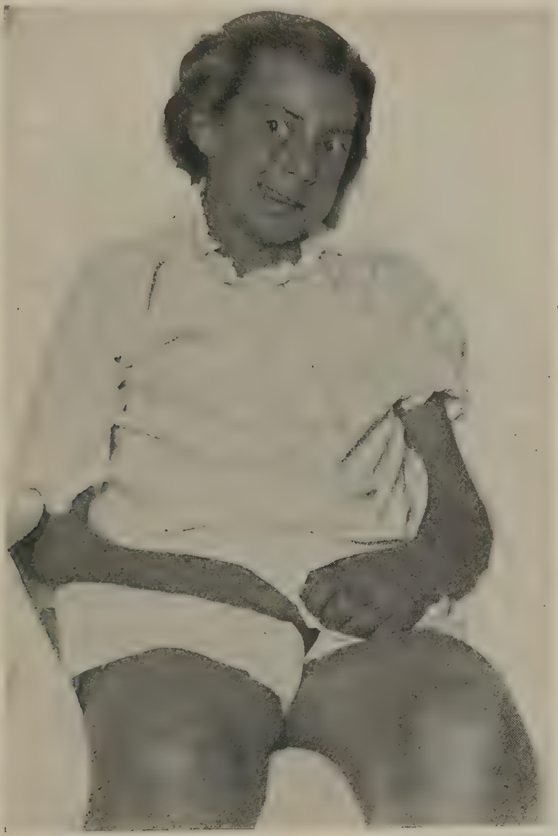

FIG. 63.—Illustration from the same case as Fig. 62 showing the left facial palsy.

The general course of the disease, which may last from six to eight weeks, leads to recovery. The cases of isolated or circumscribed paralysis recover more rapidly than the generalized forms. The ocular nerves recover very much more rapidly than do the nerves of the extremities. As a rule, from four to six months are necessary for the disappearance of the paralysis in the legs, although it may last a year or longer. The deep reflexes reappear very slowly and may be absent some time after the disappearance of other symptoms.

The older the patient, the more favorable is the prognosis. Cardiac, pharyngeal, and diaphragmatic involvement characterizes the more unfavorable cases. This is especially true of diaphragmatic paralysis occurring before the third week; after this it is not so unfavorable an omen. Very early palatal palsy indicates a probable cardiac paralysis and signifies a rapid and fatal course. The earlier the paralysis and the more quickly it reaches a marked degree, the more serious is the case. In the general form, death may be the result of inanition, asphyxiation, aspiration pneumonia, or paralysis of the vagus and phrenic nerves. Complication with nephritis or endocarditis increases the serious aspect of the case.

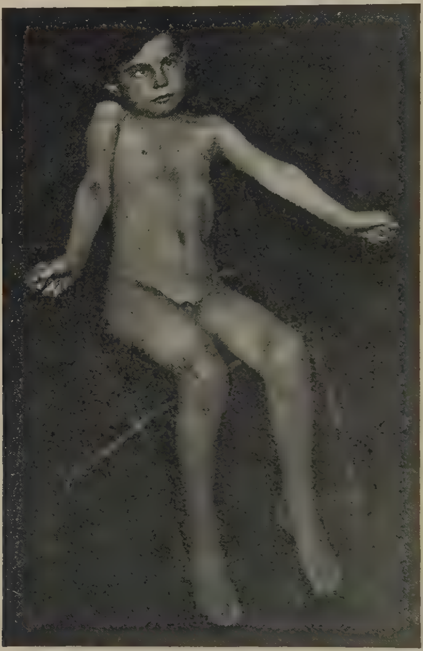

FIG. 64.—A case of multiple neuritis with no definite history of a causative factor. A history was obtained of an infection of the foot a few weeks prior to the onset of the paralysis. The case exhibits a paralysis of both upper and lower extremities, the distal portions of the lower extremities and the proximal portions of the upper being the most involved. Both facials seem weak, although no evidence of a facial diplegia other than this can be obtained.

Pathologic Anatomy and Pathogenesis.—Diphtheritic polyneuritic paralysis occurs usually following an infection of the naso-pharynx, but it has been observed in diphtheritic infection of the umbilicus, the vulva, intestines, and infected wounds on the surface of the body. The morbid changes are essentially the same as those found in other forms of multiple neuritis. The first description of the pathology of diphtheritic polyneuritis was given by Charcot and Vulpian,[49] who described degenerative changes in the nerves of a paralyzed palate. Oertel[50] found capillary hemorrhages in the cerebrospinal dura mater and in the sheaths of the peripheral nerves, and also noted infiltration about the nuclei and granulation of the anterior horns of the spinal cord. In 1879 Dejerine[51] found changes not only in the peripheral nerves, but in the cells of the anterior horns and in the anterior roots as well. An extensive literature has followed since his time. The lesions described by subsequent writers may be divided into purely muscular changes, polyneuritis, lesions of the spinal cord and cranial nuclei, and cerebral lesions due to vascular disturbances. The muscular tissue is more distinctly changed here than in any other type of paralysis due to multiple neuritis. In muscles where there has been a long standing paralysis, fatty and granular degeneration is present. Parenchymatous and interstitial changes may be found, the changes varying from simple inflammatory conditions to complete degeneration. The neuritis shows a predominance of parenchymatous change, the interstitial tissue between the nerve fibers being but little affected. The myelin and nerve filaments undergo segmentation, and the nuclei of the sheaths are increased. Abraumzellen, or granule cells, abound, marking the removal of disintegrated nerve tissue. At times, part of the nerve is involved; at other times all of it, the pathologic changes extending even to the anterior and posterior roots. The axis cylinder remains intact until considerable destruction of the medullary sheath has taken place. Thickening of the nerve is not always absent (Oppenheim); neuritis nodosa (Meyers), hemorrhages and periarteritic processes in the degenerated nerves have been observed. Although the peripheral nerves are chiefly affected, the central part of the nervous system is not immune. Lesions in the nuclei of the cord and medulla, first described by Dejerine and more recently by Rainy,[52] are found. The cells of these nuclei show various changes, chromatolysis, vacuolation, pyknosis, shrinkage, swelling, and changes in the processes. It is problematical whether these changes are primary or secondary. The probability is that they are both—primarily, as direct toxic changes, and secondarily, as the result of axonal or indirect reaction. Thickening and hemorrhages into the meninges, hemorrhage into the oculomotor nuclei, and changes in the ganglion cells of the motor cortex have been described.

In general it may be said that the pathologic changes due to diphtheria are the result of widespread action upon all of the nervous tissue, but are most marked and constant in the peripheral nerves, appearing, however, in the anterior, sometimes in the posterior horns of the spinal cord, the cranial nuclei, the motor ganglion cells of the cortex, and the meninges. The changes in the cerebrospinal fluid, increased globulin content and pleocytosis (Roemheld[53]) speak for a generalized action of the toxin.

The changes in the nervous system are more likely the result of a toxin than of direct microbial action. Experimental work (Roux and Yersin[54]) seems to indicate that if this is the case, the early appearance of palatal paralysis speaks for a direct action of the toxin upon the nerves supplying these muscles. This is made more probable by such cases as reported by Kussmaul in which an infection in the umbilicus was followed by early paralysis of the muscles of the abdominal wall. Babonneix[55] has shown experimentally that the toxin produces an ascending neuritis of the nerves to which it is carried by the lymph stream. The peripheral neuritic changes as well as the isolated paralysis of accommodation can be explained only as the result of a generally distributed intoxication by means of the circulatory system from the local infection. The predilection for certain parts of the nervous system, such as the ocular muscles of accommodation, must be emphasized.

The toxin has recently been shown by Simmons[56] to produce its changes by acting as a cell stimulant, causing a depression of functional activity.

Diagnosis.—The diagnosis of post-diphtheritic paralysis is, on the whole, not difficult. With a history of preceding diphtheria clinically and bacteriologically determined, or even an uncertified angina, the appearance of polyneuritis leads to the assumption of the presence of a diphtheritic paralysis. In the absence of such a history, the appearance of an isolated paralysis of the palate or muscles of accommodation is specific for post-diphtheritic paralysis. The severe forms, involving the vagus nuclei, the phrenic nerve, and the nerves of the cervical cord, are usual only in diphtheria and are seldom, as in poliomyelitis, found elsewhere. Juvenile tabes dorsalis is made improbable by the acute onset, the absence of pain, the absence of an Argyll Robertson pupil, of bladder and rectal disturbances, of marked objective sensory disturbances, as analgesia, of primary optic atrophy, a negative Wassermann reaction, and different serologic picture in the cerebrospinal fluid. The affection must be differentiated from poliomyelitis in cases where the degenerative paralyses are the most marked features. Typical for diphtheria and rare in poliomyelitis is the occurrence of early palatal paralysis and isolated paralysis of ocular muscles of accommodation. In poliomyelitis the onset of paralysis comes soon after the initial symptom; in diphtheria it comes late. Frequently the initial symptoms of poliomyelitis are more violent than those of diphtheria. The paralysis is usually symmetrical in polyneuritis and asymmetrical in poliomyelitis. Sensory disturbances, though they may not be marked, are prominent in polyneuritis and absent in poliomyelitis. Renewed attacks of further symptoms of paralysis speak for multiple neuritis. Involvement of the cranial nerves, which is almost never found in poliomyelitis, points toward polyneuritis.

The other conditions from which diphtheritic paralysis must be differentiated are the same as those which have been dealt with under the general subject of multiple neuritis.

Treatment.—As indicated above, it is considered probable by some authorities (Wettstein) that post-diphtheritic paralysis may be prevented by the proper employment of antitoxin. It is more or less the general opinion, however, that after paralysis has set in, antitoxin is of no great avail. On the other hand, Comby, Bruns, Babonneix, and others, maintain that it has a beneficial effect upon existing paralysis.

The treatment of diphtheritic paralysis differs in few respects from the treatment of polyneuritis in general. Mild cases of palatal palsy or accommodation paralysis require no treatment. The liability to heart complications makes it imperative to keep the patient in bed until all danger is over. Rolleston observes that sudden death due to cardiac failure is not the bugbear it is usually considered by practitioners. Careful observation of the function of the heart should be made, and the recumbent position insisted upon at the slightest indication of involvement. It seems unnecessary to insist that there should be an abundance of fresh air. The strength of the child must be supported by food of a nourishing and digestible character. Special attention must be paid to the possibility of a complicating nephritis. When, as the result of vagus involvement, dysphagia is present, early feeding per rectum or by the stomach tube should be instituted. Milk, eggs, broths, flour soup, or a combination of these with sugar may be given four times a day by means of the stomach tube, special care being taken that the tube is passed into the stomach, inasmuch as the palatal palsy and anesthesia, along with the anesthesia of the larynx, makes it very easy to pass a nasal tube into the trachea. For this reason, semi-solid food may at times be given when fluids should be interdicted. When it is advisable, rectal feeding of predigested foods or glucose, or both, should be used to prevent inanition and subsequent death. Small pieces of ice may be placed in the mouth to slake thirst. If paralysis of the diaphragm occurs, the child should lie with the body partly raised and supported by pillows. Small doses of strychnia should be used, and faradization of the diaphragm may be employed. Only in the absence of involvement of the heart are diaphoretic measures permissible. The administration of strychnia and tincture of nux vomica is serviceable. Rolleston advises adrenalin by mouth every two hours, to be used as a prophylactic measure. This treatment may be continued after the development of the paralysis. When the paralysis has passed the acute stage, the methods employed in the treatment of any polyneuritis are indicated. Massage, passive movements, graduated exercises, electrical treatment, and reeducational methods may be employed. For paralysis of the muscles of deglutition, one should apply galvanism to the muscles of the throat, making a stroking motion downward and eliciting the swallowing reflex.

Other Forms of Infectious Neuritis.—Multiple neuritis occurring as the result of typhoid fever manifests itself either during or subsequent to the pyrexial period of the disease. The paralysis is of the amyotrophic type; it is usually partial in extent and degree and is associated with reaction of degeneration. It is almost invariably preceded or accompanied by some form of sensory disturbance.

Of the paralyses following scarlet fever, Bury says that no cases known to him (up to 1892) could be attributed with certainty to a peripheral neuritis. Basette reported a case of a child 10 years old with paralysis of the upper and lower limbs, with atrophy and deformities which followed scarlet fever. He said that the paralysis was in all probability the result of an inflammatory infection which attacked both nerves and cord and was due to the action of the infection upon the peripheral nerves and the nerve centers.

Some of the cases of paralysis following measles are probably of peripheral origin. Denarié mentions several cases, in some of which the clinical description affords evidence of the possible presence of peripheral neuritis.

Beriberi.—Although adult beriberi has been reported from many quarters of the world, infantile beriberi has been restricted to few countries, chiefly Japan and the Philippine Islands. In 1888 Hirotu[57] described this condition, which he called infantile beriberi. Later a Filipino, Guerrero,[58] described a clinically similar condition among Filipino infants. More recently it has been described by McLaughlin[59] Andrews,[60] Chamberlain,[61] and Vedder.[62]

Infantile beriberi or kakké causes practically 50 per cent. of the deaths in the Filipino infants under one year of age. Of these 75 per cent. are breast fed. It is practically always associated with adult beriberi in the mother.

The onset of the disease is insidious. The baby has a round face and appears well nourished. The early symptoms are indicated merely by a slight edema of the legs, which manifest tenderness on pressure and pain on movement. The knee-jerks are usually absent.

Alcoholic Multiple Neuritis.—Alcoholic multiple neuritis is rarely observed in children. Campbell[63] in 1893 described a case in a child five and a half years old. The history, however, was not very clear as to the alcoholic features. Lescynski,[64] in 1892, described a case in a child seven years old in which amyotrophic processes and disturbances of sensation were present. Lyon Gaston reported a case in 1887. In 1893, Jacob[65] reported a typical case in a boy five and a half years old. Since then Zappert[66] speaks of a similar case, Cassirer[67] reports a case in a boy three years old, Wallfield[68] a case in a boy of seven, Herter[69] a case three years old, and Eichorst[70] one eight years old. Zappert believes that multiple neuritis occurs in children as the result of their consuming large quantities of beer. In Campbell's case, however, spiritous liquors were the cause.

Remak and Bruns describe a form of alcoholic neuritis in which, at different times, various nerves are involved asymmetrically in a mononeuritis. This condition Remak terms mononeuritis multiplex, or disseminated mononeuritis.

The symptomatology of alcoholic multiple neuritis differs in but few respects from that of the adult type. The amyotrophic form is most common, although the ataxic form has been observed. Usually the disease is ushered in by severe pain and attains its development rapidly. As in the adult form, neuralgic pains are very severe. The muscles are tender to pressure and are frequently hard in consistency. They are often the site of a myositis. Hyperesthesia of the skin and edema is present in many cases. The reflex functions of the bladder and rectum are intact. Delirium, convulsions and coma may be associated with multiple neuritis although, as a general rule, these conditions may be traced even in childhood to excessive alcoholic indulgence. Korsakoff's psychosis has been observed in a case by Sereni. Although a fatal case has been reported, the prognosis on the whole is good. The prophylaxis of this condition, namely, the prohibition of the use of alcohol for children, is seemingly a procedure needing no particular emphasis, even to the lay public. Nevertheless, in Jacob's case, the father stated that "the child was always thirsty, and of course we could not give it water."

The treatment of alcoholic multiple neuritis, aside from the removal of the etiologic factor, is the same as that employed for any of the multiple neuritides.

Lead Multiple Neuritis.—Peripheral lead palsy or polyneuritis is far more common in children than alcoholic polyneuritis. In 1892 Lockardt, Gibson, Love, Turner, and others[71] drew attention to a disease, which they attributed to lead poisoning, occurring amongst children living in the neighborhood of Brisbane, Australia. Since that time further contributions have appeared on several occasions [72,73] from the hands of these men. Bruns, in 1912, reported 40 cases in the literature, described by Putnam,[74] Sachs, Hahn, and Oppenheim. Since then a number of other cases have been recorded.[75,76] The entrance of lead into the body is most likely to occur in the children of parents who are employed in the lead industries. The poisoning, on the other hand, may be the result of lead derived from glazed fruits, acid fruits, vegetables put up in tins, candy wrapped in tinfoil, certain rubber nipples, various kinds of cakes and candies which are treated with lead, food stuffs cooked in utensils which are carelessly soldered, and the inhalation of lead from freshly painted walls. A great source of danger in rural districts is the drinking of water conducted through lead pipes, either connected with a pump or conveyed into the house. The use of leaden playthings, such as trumpets, etc., may be responsible for the disease. Raudnitz and Holm have pointed out that lead plasters, Hebra's ointment, etc., may be the cause in some cases. Finally, Anker and Oppenheim[77] reported a case of lead palsy in a child in whom it was thought the condition was hereditary.

The symptoms of lead palsy in children are similar to those in adults. Turner[78] divides the cases into four groups: (1) Paralytic cases showing symmetrical wrist-drop and foot-drop, spasms of calf muscles and, as a secondary lesion, a permanent talipes equinus. (2) Cases characterized by pain in the abdomen and limbs, concomitant with habitual constipation and an occasional blue line of the gums, the children becoming at the same time irritable and neurotic. (3) Eclamptic cases suffering from severe and persistent convulsions which often end fatally. (4) Children suffering from ocular neuritis accompanied frequently by a neuritis of the oculomotor nerves.

As a rule, the onset of the paralysis is preceded by other symptoms of lead poisoning; lead colic, sometimes pains in the joints or muscles, or by symptoms of brain disease, such as lead encephalitis. Often there is found a lead line or blue line at the margin of the gums close to the teeth. Anemia may be present and at times cachexia. The lead palsy, however, may be the first and only symptom of the intoxication. It is notable that in contradistinction to the adult form, the lower extremities are first involved and later, the arms. Bruns has suggested that this difference may be due to the fact that children, in contrast to adults, use their lower extremities more than the upper. The legs and feet, therefore, are the first to be affected since the localization of lead paralysis is influenced to a considerable degree by overstrain of muscles. The peroneal group of muscles is the chief site of trouble in the lower extremities. At times the small foot muscles may likewise be involved. In the upper extremities the muscles chiefly affected are those supplied by the musculospiral nerve, the extensors of the fingers and hand, and later of the thumb. The abductor of the thumb and the supinators, especially the longus, and the triceps are not involved. If the paralysis is very slight, it may be limited to the extensor communis digitorum, or to some branches of these muscles. Usually the wrists are flexed, as a result of which the fingers seem weaker, but if the hand be passively moved to the normal position, flexion becomes as strong as usual. At times the small hand muscles are involved in the paralysis, most frequently the muscles of the thenar eminence; at times the deltoid may be included.

The paralysis is flaccid, with atrophy and reaction of degeneration There is neither any pain nor any disturbances of sensation. The deep reflexes are absent in the zone of paralysis. The visceral reflexes are normal.

The morbid anatomy of lead palsy, according to Gombault,[79] shows a periaxillary neuritis, the sheath of the nerve being more affected than the axis cylinder. Although the greater force of its action is spent upon the peripheral nerves, the cord as well as the brain may be affected.

The diagnosis is relatively easy. The history of exposure to intoxication, the signs of lead palsy, and the peculiar type of paralysis comprise a typical picture.

The prognosis is good when further poisoning can be avoided. When the paralysis has recurred repeatedly the prognosis is more serious.

The treatment may include measures which increase the elimination of lead, as diaphoresis, catharsis, and administration of the iodides. Lockardt[80] recommends lumbar puncture in all cases of lead poisoning in which there are eye symptoms. Aside from this, the treatment is the same as for the other neuritides.

Arsenical Multiple Neuritis.—Arsenical multiple neuritis is most frequently the result of medication, usually from the use of Fowler's solution in chorea (Brouardel,[81] Raymond,[82] Colman,[83] Karplus,[84] Kron[85]). It may result, of course, from such vermin destroying mixtures as "Rough on Rats" and German fly paper. Furthermore, arsenic is contained in many of the covers used for books, in toys, in labels, as well as in artificial flowers.

It manifests itself as an amyotrophic paralysis in the arms and legs. The affection develops rapidly; pain and paresthesias are very prominent. Usually the distal segments of the extremities and the extensors (peronei, musculospiral) are chiefly affected. According to Oppenheim, however, the paralysis affects the small muscles of the hand much oftener in arsenical than in alcohol or lead polyneuritis. The paralysis is an atrophic one; reaction of degeneration is present. The deep reflexes are absent; the muscles and nerves are tender to pressure; anesthesia and hyperesthesia are common over the legs and feet and the hands and feet. Bladder disturbances may occur. Ataxia is as common as in the adult form. Trophic disturbances are usually found. Herpes zoster, pigmentation, glossy skin, loss of hair, and hyperkeratosis of the hands and feet are observed.

The prognosis on the whole is favorable, but the convalescence may be protracted over months or years. A few cases of secondary contractures have developed in the antagonists of the paralyzed muscles.

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THE NEURALGIAS

The term, neuralgia, is used to designate a condition characterized by pain in the course of a nerve or its branches. It comes on in attacks of paroxysms, lasting from a few seconds to minutes, and consists of groups of repeated single pains which are quite severe in nature. In the interval between the attacks, pain or other disturbances are completely absent, except occasionally for a persisting, dull sensation. The attack itself represents an exacerbation of this sensation. With very few exceptions, such as trifacial neuralgia, for example, the neuralgias are only symptoms, not distinct diseases. The recent trend of thought is to do away with the term in such cases, using it only to indicate a nerve pain. It is a term usually employed for those cases in which we have persistent nerve pain, and for which we can find no organic basis.

Excluded from the neuralgias are all cases of neuralgiaform pains occurring in the course of organic nervous disease, as in tabes, syphilitic meningitis, tumors of the brain and spinal cord, herpes zoster, such cases of injury of the nerves and traction and compression by cicatrices, which cause a neuritis, degenerative disease of the nerves themselves, neuralgiaform pains due to arthritis, bursitis, osteitis and periostitis, and malformations causing pressure upon nerves as cervical ribs, and enlarged transverse spinous processes of the lumbar vertebræ. In its strictest sense neuralgia is rare in adults and particularly rare in children.

The neuralgias are classified according to locality and cause, such as hysterical, gouty, trigeminal, cervical, brachial, etc. Of the neuralgias occurring in childhood, mention is made of sciatic neuralgia (Bruns),[1] occipital (Zappert),[2] and trifacial (Bruns). Many of the cases of face pain in children which have been described as trifacial neuralgia are really due to sinus disease or other infective or mechanical diseases of the head. Real trifacial neuralgia, however, does occur. Of 200 cases of trifacial neuralgia reported by Patrick,[3] only one began in childhood and that at the age of seven. Another case, which he reports but which is not included in this series, began at eight. Trifacial neuralgia, which is short-lived, nearly always begins as an instantaneous sharp pain, rarely lasting longer than 10 to 40 seconds. Paroxysms of succeeding pains, however, may last an hour. The pain is severe and usually occurs spontaneously, although it may be started by relatively slight peripheral irritation. The area which starts the pain, when irritated in this manner, has been called by Patrick the dolorgenetic zone. It is not always affected by the pain which it elicits. For example, the "Trigger zone," as it is called, may be in the distribution of one division of the nerve and the pain in another. Eating, talking, swallowing, may determine the irritation of the trigger zone as well as rubbing or touching the face.

The treatment of the neuralgias, aside from trifacial neuralgia, is purely symptomatic, inasmuch as these conditions are classified as neuralgias only because we are ignorant of their cause. Trifacial neuralgia in the child is treated in the same manner as in the adult. Palliative treatment consists of alcohol injections or resections of the peripheral nerves, and radical treatment by section of the sensory root of the Gasserian ganglion or removal of this ganglion.

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HERPES ZOSTER (SHINGLES)

Herpes zoster is a name applied to an eruption of vesicles, following the distribution of one or more nerves, corresponding in extent, either to the areas represented by the posterior roots or, according to another theory, to the sensory cutaneous areas of the spinal segments. The eruption most frequently develops on the trunk in the region of the dorsal roots. The name, herpes zoster, was chosen from the fact that their distribution usually extends in the form of a half girdle. The trigeminal area is frequently affected, chiefly in its ophthalmic branch. The condition is named according to its distribution: zoster occipitocollaris, cervico-brachialis, dorso-abdominalis, lumbo-inguinalis, sacro ischial; or according to Head, herpes sterno-nuchialis, cervico-subclavicularis, dorso-ulnaris, etc. In addition to the well-known clinical picture of involvement of the spinal and Gasserian ganglia, the ganglia of the other cranial nerves may be affected, as the seventh, geniculate ganglia; the ninth, petrosum; and Ehrenritter; the vagus; jugulare and plexiforme (Hunt).[1] There is strong reason to believe that the auditory ganglia may be affected likewise (ganglia of Corti and Scarpa). The syndrome of the geniculate ganglia is a herpes zoster oticus alone or accompanied by facial and auditory complications.

Herpes zoster may be a primary or a secondary affection. In the latter case, it may be caused by injury affecting either the nerve or its roots or the spinal ganglia. The observations and investigations of Head and Campbell,[2] however, show that it is frequently an independent affection, a primary, acute, infective disease. Some of the factors substantiating this belief are the febrile character of the illness, the swelling of the glands, and the immunizing influence of the disease. Microörganisms have been found in the cerebrospinal fluid (Achard and Loper, Widal, Brissaud, Sicard). It has been observed to appear in epidemic form (Rohe, Kaposi, Weis, Reilly, Dopter, Sachs). It has been noted in the course of other infectious diseases.

Poisoning with arsenic (Hutchinson,[3] Gerhardt,[4] and with carbon monoxid (Leudet[5]) may produce herpes zoster.

Sunde,[6] in a careful examination of a Gasserian ganglion of a patient 87 years old, found an acute inflammation with hyperemia, hemorrhages, and round-celled infiltration. Barrensprung[7] was the first to show that herpes zoster may result from a disease of the spinal ganglia. Head and Campbell stated that underlying a primary spontaneous herpes there is often an acute, frequently hemorrhagic inflammation of the spinal ganglia. Rosenow and Oftdal[8] produced herpes zoster of the tongue and lips and lesions in the corresponding ganglia in animals by intravenous injections of bacteria found in emulsions of extirpated tonsils or from pyorrheal pockets. Cocci were found in the ganglia by cultures and sections, even when they were absent from the blood and other tissues. On the other hand, Curschman and Eisenlohr proved that in some cases the disease was neuritic in nature (a neuritis nodosa). Changes in the spinal cord and hemorrhagic foci in the gray matter of the corresponding lateral horn and degeneration of the communicating rami of the sympathetic nerves was found in a case by Thomas.[9] In the great majority of cases, the disease is limited to one side, but a bilateral distribution has been observed in a few cases (Kaposi, Thomas, Weis, Hartzell). In childhood the simultaneous occurrence of herpes and poliomyelitis is rare. One case is quoted by Batten,[10] who says that nearly all the recorded cases are in adults. It seems certain that many of these cannot be attributed to the virus of poliomyelitis.

Herpes zoster is frequently a febrile disease, the eruption being accompanied by fever and its attendant symptoms. A rise of temperature may precede the vesicles. Swelling of the lymph glands is common in this form. The vesicles appear in groups upon a background of reddened, perhaps swollen skin. The contents, at first composed of clear serum, become opaque and purulent, seldom hemorrhagic. They dry up and are replaced by brownish crusts or scabs which desquamate leaving slightly pigmented spots. At times the vesicles become confluent and form large bullæ. While the eruption tends to disappear in a few days, the whole process may extend over weeks or months.

Neuralgia is one of the most frequent and almost constant accompanying symptoms. At times it precedes the eruption and may long outlast it, especially in old people. The pains occur in groups of sharp paroxysms, which are at times of a burning character, upon a background of a dull ache or numbness. Pruritis, loss of hair, disturbance of sweat secretion, and anesthesia frequently appear in the affected area. In ophthalmic zoster, with anesthesia of the cornea and conjunctiva, neuroparalytic keratitis may develop. Extension to other nerves from the affected ganglion is at times observed as a facial paralysis occurring in the course of a trifacial or cervical zoster (Ebstein, Frazer).[11]

The prognosis of primary herpes is favorable. Complications make the prognosis more grave. At times the condition may even endanger life. Symptomatic zoster runs a course dependent upon the primary disease. Neuralgia may be persistent, and according to Petren and Bergmark,[12] is to be expected as long as the sensory disturbances are marked.

The treatment of the vesicles consists in protecting them from rubbing and irritation. Cocain ointment is frequently of service.

The neuralgia should be treated as any other neuralgia. Epidural injections of soothing drugs have been employed. Lumbar puncture with the removal of 20 c.c. of fluid has had a curative action. Persistent and severe trifacial pain following herpes zoster is at times relieved by alcohol injection into the peripheral nerves or the Gasserian ganglion.

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GROWING PAINS

In growing children, especially girls, from 2 to 10 years of age and often about the age of puberty, weakness, stiffness, and pains in the muscles are occasionally observed. Usually they occur at night or towards evening. When they come on during the night, the child is awakened by pains in the thighs or legs, so severe that they cause it to cry out. After a time the pain subsides, and the child again falls asleep, frequently to be awakened again by the pain either in the same place or in another extremity. Finally the pain disappears, and the next morning the child is as well as ever. These pains recur at varying intervals, about once a week or less frequently. They rarely affect the arms. The changes incident to growth in the upper epiphysis of the tibia and the lower epiphysis of the femur have been suggested as an etiologic factor.

Many of the so-called growing pains are in reality cases of bone disease, affections of the periosteum, joints, or muscles. On the other hand, they may be cases of rheumatism which are frequently overlooked because of the misinterpretation of the accompanying pain. As indicated by Jacobi, it is a matter of considerable importance that careful attention be paid to the study of these cases of so-called growing pains, lest any other serious organic change be overlooked, such as, for example, the onset of an endocarditis following an acute rheumatism.

In the treatment of the pains themselves, massage occupies the chief place symptomatically; whereas a search for a form of infection, its eradication, and suitable treatment for such organic conditions as may be masked by these pains comprises the rational treatment.

SPASMS

Clonic or tonic muscular spasms which are due to organic change in the peripheral spinal nerves or muscles occur very rarely in children. Bruns emphasizes the necessity for excluding all tonic and clonic spasms, such as hysteria and epilepsy, which develop on the basis of a neurosis. Myoclonus multiplex should likewise be excluded. The head shaking, jerking, nodding, rotating movements of the muscles of the neck occurring in nodding spasm, or spasmus nutans, associated with nystagmus, should be excluded as being a clinical entity. Although chorea minor is not a neurosis, its pathology is not limited to the peripheral nervous system. Athetoid movements, spasmus mobile accompanying cerebral diplegia or hemiplegia, and reflex muscular spasms resulting from meningitis or caries of the spine, do not belong in this classification. Tetanus and tetany, general diseases showing special disturbances in the nervous system and along with them laryngospasm, must likewise be considered elsewhere. Secondary contractures and ischemic contractures must be excluded. Clonic spasms of the diaphragm, singultus or hiccough, when of an organic nature, are at times due to disease of the brain stem. In the region of the shoulder and neck, spasms may occur on rheumatic or neuritic basis. They may affect the rhomboids, the levator scapulæ, or both. At times the trapezius is affected. Spasm of the levator scapulæ has been observed following meningitis (Hughes, Bernhardt). Many clonic spasms occurring in children have been examples of tics or hysteria. Torticollis, as observed in children, where the condition is not due to a spasm but to a deformity or a congenital absence of some of the muscles may either be congenital or reflex in character, consequent to disease of the cervical vertebræ.

Muscular cramps, occurring usually in the calves of the legs, are frequently observed in children. They often follow some unusual exertion, with the first movements of the legs following a rest; thus they occur in the morning while the child is in bed. They consist of sudden, strong contraction of a mass of muscle with a resulting movement of a segment of the extremity, associated with severe pain. They last from a few seconds to several minutes and disappear spontaneously. Their disappearance is frequently hastened by passive change of position of the segments involved in the cramps.

The treatment of the various spasms is dependent upon their cause. Massage, exercise, and thermal measures occupy a prominent place in the treatment of the spasm or cramp itself.

POLYMYOSITIS

Polymyositis has been divided by Lorenz[1] into several forms: (1) Dermatomyositis; (2) polymyositis hemorrhagica, which usually follows tonsillitis and may be associated with myocardial involvement. It resembles acute dermatomyositis, but is characterized by the formation of hemorrhagic foci between the muscles. The disease often ends fatally. (3) Polymyositis with erythema multiforme. The cutaneous affection presents the usual appearances of erythema multiforme and on careful examination painful nodules in the muscles can be felt. The disease is usually benign. (4) Neuromyositis.

In children only dermatomyositis has been observed.

In 1887 polymyositis was described as a clinical entity nearly simultaneously by Wagner,[2] Hepp[3] and Unverricht.[4] The latter pointed out the constant accompanying disturbance of the skin and designated one group of this condition as dermatomyositis. Shortly after a number of authors also described the condition. In 1898 Lorenz collected 15 cases. Schüller,[5] in 1903, by adding his case of a boy seven years old, brought the number of cases in children to five. Since then a number of other instances of the disease in children have been reported (Bruns, Oppenheim).[6] In 1903 Steiner[7] collected 28 cases among adults and children.

Dermatomyositis is a clinical entity of acute, subacute, or chronic course, of unknown origin, characterized generally by a gradual onset, with vague and indefinite prodromes, followed by edema, dermatitis, and a multiple muscle inflammation.

The disease generally attacks persons in the prime of life, although Fuckel's[6] case was noted in a girl after an attack of measles. Both of Wagner's[9] cases had pulmonary tuberculosis, and Senator's second case was a diabetic. The onset of the disease is almost always gradual. It may, however, be sudden with the prodromal symptoms of malaise, weakness, pains in the extremities, headache, anorexia, and, especially in children, gastric disturbances. These symptoms may last from several days to three weeks. On the other hand they may be entirely absent. Fever is soon noted, usually of moderate intensity and intermittent or remittent in type, and rarely exceeding 104°F. Not infrequently an enlargement of the spleen occurs. Vague pains and a stiffness or rigidity in the extremities and back form the next step in the attack. Movements become difficult and walking painful, so that in a short time the patient is confined to bed. The pains are described as drawing, tearing, and boring. They rapidly take on a more definite character and become circumscribed or localized, different muscular groups being successively attacked. Eventually the whole body is affected. The pains occur not only spontaneously, but also upon active and passive movements. In most cases the patient finally lies in bed utterly helpless as if he were completely paralyzed. The muscles are very painful to pressure. At times they are swollen and hard and at other times soft. At times contractures have been observed. Frequently the edema conceals the condition of the underlying muscles. As the disease progresses, the muscles may become atrophic. Because of the severe pain and the edema of the skin, electrical examinations have been few and unsatisfactory. Oppenheim[10] states that there is generally a quantitative decrease in the electric excitability of the muscle, and Lewy[11] obtained a partial reaction of degeneration in his case. As a rule, the nerves are not tender to palpation. Sensory disturbances are rare; Wagner, Lewy, Lorenz, and Leube have noticed paresthesias during the course of the disease. The knee-jerks in the mild cases may be slightly increased; in the moderate cases normal, and in several very severe cases they were absent. The muscles of the proximal parts of the extremities are more frequently involved. The hands and feet are often unaffected. The muscles of the neck, back, and abdomen may be implicated as well as the muscles of respiration, the intercostals, and the diaphragm, as the result of which asphyxia may ensue. The process may extend to the muscles of deglutition, so that aspiration pneumonia may result. The muscles of the face and the eyes may likewise be included in the disease. The heart muscle is unaffected.

The second characteristic symptom is the dermatitis and edema, the latter appearing with the fever and at times involving the whole body. Generally it is first seen on the face, especially above the eyelids, and may finally cause the face to assume an aspect likened by Oppenheim to alabaster. The edema is more frequently present in the proximal parts of the extremities, the shoulder, arm, elbow and thigh. It is asymmetrical. At times it may pit on pressure, but usually a dense, hard infiltration of the subcutaneous tissues is observed, the duration of which is usually limited to the acute stage of the disease.

The dermatitis may occur in the form of an erythema, at times resembling erysipelas, an urticaria, a roseola, an eczema, or an inflammation resembling erythema nodosum. It appears not only on the extremities, but also on the trunk and face. Usually it is present over the affected muscles. It may occur only later in the disease. Frequently it disappears without leaving a trace, but at times some pigmentation results. Stomatitis and angina may be present. In Lewy's case and in four of Oppenheim's, ulceration of the mucous membrane occurred, as a consequence of which Oppenheim termed these cases dermato-mucoso-myositis. Profuse perspiration frequently accompanies the other symptoms.

The acute cases run their course in one to eight weeks. When death occurs it is usually the result of an aspiration pneumonia. The subacute cases occupy a period of from two and one-half to six months, and the chronic from one and one-half to two years. In these subacute and chronic cases there is either a continuation of existing symptoms or exacerbation of symptoms in other groups of muscles one after the other, each associated with a febrile disturbance. Frequently in the chronic forms muscular atrophy ensues.

Diagnosis.—As a rule no difficulty will be experienced in the diagnosis of this condition. In trichinosis the initial gastro-intestinal disturbances, the discovery of the trichina in the stool, the excised muscles, and the cerebrospinal fluid determines the diagnosis. In neuromyositis, the absence of dermatitis and the presence of neuritis symptoms, actual paralysis, reaction of degeneration, and sensory changes are indicative of this condition. Septic, pyemic, and syphilitic polymyositis must be excluded. One of Oppenheim's patients turned out four years later to be a typical example of scleroderma.

Prognosis.—Of 28 cases collected by Steiner, 17 terminated fatally. The prognosis depends upon the extent of involvement of the disease. If all of the muscles are involved, including those of respiration and deglutition, death probably will be the result.

Pathology.—The skin covering the involved muscles is firm and hard. The subcutaneous tissue presents a firm and tense edema and is infiltrated with a yellowish serous fluid. The muscles are swollen; they are pale red or yellow in color; and may occasionally reveal a yellow-gray streak. They are suffused with serum and quite moist. At times they are hard and firm; at other times soft and boggy; frequently they are friable. Microscopically, the changes are those of parenchymatous and interstitial inflammation. The fibers are found in all stages of degeneration. They may be normal in size, edematous, or atrophied; granular, hyaline, waxy, or fatty. Typical interstitial foci of small round cells are found in the perivascular connective tissue and to a lesser extent in the muscles. In the subacute and chronic cases there is an increase in the connective tissue, which may be marked in both the perimysium, externum and internum. In patients dying soon after the development of the disease, no degenerative changes were seen in the muscles, so that it would seem that the interstitial changes are the primary ones and the muscle degeneration secondary. The nervous system is intact.

The treatment is directed chiefly to the alleviation of pain and the support of nutrition. Oppenheim advises, as in multiple neuritis, the use of diaphoresis.

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PROGRESSIVE MYOSITIS OSSIFICANS

Progressive myositis ossificans is a progressive, inflammatory affection of the motor system, of unknown origin, characterized by the formation of bony masses in the muscles, aponeuroses, tendons, ligaments, and bones, with resulting ankylosis of most of the articulations.

The disease was first described by Freke[1] in 1740. The same journal also contains a description of a case by Copping. Other cases were noted by Henry, 1759; Rogers, 1830; Testelin and Dampressi, 1844; Hawkins, Wilkinson, and Abernethy. It was not however, until Munchmeyer's[2] paper appeared in 1869 that the disease attracted any notice. In this paper Munchmeyer carefully reported a case of his own and collected 12 from the literature. At DeDusch's suggestion, he adopted for the disease the designation, "Progressive Myositis Ossificans." Some years later, Helferich[3] noted the occurrence of a curious malformation in connection with this disease, microdactyly, ankylosis of the phalanges of the thumb, and a supposed absence of one phalanx of the great toe on both sides. Since his time the same anomaly has apparently been noted in 75 per cent. of the cases recorded. In 1884 Pinter[4] carefully reviewed the literature reporting 22 cases. In 1896 Pincus[5] tabulated 38 cases. Roth,[6] in 1898, in his review of the literature, mentions having found 40 cases, including his own, which number, however, includes the four cases of localized myositis reported by Cahen and Schwarz. In 1890 De Witt[7] in a careful search of the literature, brought the number of cases to 78. In 1908 Walker[8] collected 92 cases, and Elliot[9] in 1911 by adding one of his cases collected 107.

Progressive myositis ossificans is a rare disease, one which is typical of young people, as Lorenz's records in his monograph on muscle diseases well show. Of 45 cases in which the age at onset is given, 38 were affected before the fifteenth year. Eleven of these showed the disease during the first year of life, 16 between the first and fifth years, and 11 between the ages of 5 and 15. It occasionally happens that the patient does not fall a victim to the disease until later in life. Rogers' patient was 35 years old when stricken. In 1898 Weil and Nissin[10] collected 42 cases in children, 15 of which began in the first and second year, 17 from the second to the fifth year, and 10 from the fifth to the fifteenth year.

From 1899 to 1909 Bruns[11] found nine cases in the Jarhbuch f. Neurol. u. Psych. These cases occurred in boys and girls ranging in age from two to nine and a half years. The disease occurs much more frequently in boys than in girls. Gerber[12] gives the ratio as 4:1, while Rolleston[13] says it is 5:1. In Weil and Nissin's series, 38 were males and 12 were females. Pincus tries to account for this difference by laying particular stress on obstetrical traumatism. He attributes the preponderance of male cases to the more frequent injuries of boys at birth, due to their better development.

The etiology is not known. Hereditary taint plays no rôle. One must be content with the vague hypothesis that predisposition is due to some aberration of growth congenitally acquired. Some claim that this aberration comes from an ossifying predisposition, and that persons possessing it may develop the disease from some exciting cause, such as cold, insanitary surroundings, trauma, either slight or severe, and single or successive injuries. It may occur from no known factor.

Mays,[14] Ziegler,[15] Lorenz,[16] and other investigators favor this view of ossifying predisposition. Other theories suppose it to be a trophic neurosis, a change similar to progressive muscular atrophy and hypertrophy, a rheumatic condition allied to muscular rheumatism and rheumatoid arthritis, and a reversion to a later type due to atavistic influences.

There are at present two main theories. The first considers the process of bone formation to be of an inflammatory nature, while the second view regards the bony growth as representing a tumor formation. The present trend of opinion appears to be towards the second view. Rolleston and Elliot, however, adhere to the former theory, stating that myositis depends first on a congenital weakness and, second, on a tendency to aberrant growth on the part of the mesoblast. As the result of diminished resistance, the muscles are more susceptible to involvement, while the tendency to aberrant growth subsequently shows itself in a calcification and ossification of the inflammatory products. The correctness of the term myositis is very doubtful. Goto,[17] in 1913, suggested the substitution of the name "Hyperplasia Facialis Ossificans Progressiva."

Weber and Compton[18] state that the disease, which advances by repeated fits and starts, appears histologically to be an ossifying hyperplasia of the fascia and connective tissue of the striped muscles, especially the muscles of the trunk. They think that the microscopic examination of the diseased muscle, before any local bone formation had occurred, showed in their case that myositis, in the strict sense of the word of an ordinary inflammation, has little to do with the disease. That a predisposition is necessary for the development of this disease is shown by the fact that it affects the youngest children, that it frequently is symmetrical, and is associated with developmental defects, as microdactyly.

Symptoms.—The disease develops insidiously and may exist for years before it is recognized. In fact, many cases are diagnosed as rheumatism, their true nature being revealed only at the period of bone formation. In typical cases the signs of a myositis are present without any known exciting cause. Sometimes, after trauma which may be very insignificant, a swelling presenting a firm, doughy consistency is found in the involved muscle. Pain may be present, either confined to the muscle attacked, or radiating in character. The overlying tissues are edematous, and the skin shows some redness. Usually the temperature is slightly elevated. In the course of a few days, depending upon the intensity of the process, these symptoms vanish, but the muscle does not return to its normal state. It remains indurated, and finally a hard, tumor-like mass develops. The disease at this point may become stationary for some time. Eventually, other masses are formed, some forming bony deposits, some remaining as fibrous tumors, and some disappearing, causing the muscles to undergo fibrous degeneration. Subsequently, either from trauma, or from some unknown cause, the disease progresses, more muscles become involved, and bony changes may occur which are usually painless. The muscles of the back and neck are generally the first involved. They were the first implicated in 21 out of 38 cases reported by Lorenz. In other instances, the muscles of the upper and lower extremities and of the face were the first attacked. The ossification of the cervical ligaments and the muscles of the neck causes the head to be bent forward and the neck to be fixed. The ossification of ligaments, fasciæ, and muscles of the back varies to a marked degree. Very often the muscles are firmly attached to the skeleton. General bony foci may appear in many of the muscles of the back, like the ramifications of coral or a geographic map of a mountainous region. The vertebral column eventually becomes fixed, and a scoliosis is frequently noted. A kyphosis is more rare. The scapulæ early become attached to each other and to the thorax, while occasionally they may be connected with the ilium. The movements of both arms are very much limited. If the pectorals and latissimi dorsi are also involved, the arms are rigidly fixed on the thorax, which is described as a bony coat of mail. The upper extremities are usually attacked earlier in the course of the disease than the lower. The arms are generally fixed at the shoulder, and by ossification of the biceps, are flexed at the elbow. The position of the hip joints, as well as the ossification of the gluteal muscles, causes the hips to be slightly flexed upon the pelvis. This posture displaces the patient's center of gravity and gives him a precipitous gait. The forearms, hands, legs, and feet are rarely involved. At times the temporals, masseters, and pterygoids are attacked so that the patient is unable to use his lower jaw. The other facial muscles, the muscles of the tongue, eye, larynx, diaphragm, heart, perineum, and sphincters are unaffected. The process of bony formation occupies no fixed time; generally it lasts from two to eight months. The presence of exostoses has frequently been described. They are usually seen on the internal surface of the arm, the anterior aspect of the tibia, the upper part of the fibula, the ribs, or where the long bones approach the skin. The electrical excitability of the muscles varies. Polar change has been observed. Pinter found reaction of degeneration.

Microdactyly is present in 75 per cent. of the cases. The toes and thumbs are much shorter than normal, due to atrophy of the metatarsal and metacarpal bones, and subsequent ankylosis of the interphalangeal joints. This gave the impression, before the introduction of the *x*-rays, that one phalanx was missing. The great toes are generally directed outward and frequently lie under the second toe, giving rise to the deformity of hallus valgus.

Other malformations, such as the lack of some muscles, and atrophy of the testicles and scrotum, have been noted in individual cases.

Prognosis.—Progressive myositis ossificans is a condition which requires for its full development a number of years. Relapses alternate with pauses in its progress. At times there appears to be a cessation in the symptoms for 6, 10, or even 23 years, but it is a progressive disease in which the slightest cause may bring about a relapse. Upon the whole, the condition is progressive, so that finally the patients become practically helpless and bedridden. Death may ensue from pyemia as the result of bed sores and abscesses about the bony deposits, from inanition, or from some intercurrent disease, such as pulmonary tuberculosis or pneumonia. Because of the ossification of the muscles of the chest, lung affections are very prone to develop; suffocation may intervene, owing to limitation of costal breathing and edema of the glottis.

Pathology.—Munchmeyer's classification, which has been closely followed by other writers, recognized three stages in the disease. (1) Stage of embryonic infiltration; (2) stage of connective tissue induration; (3) stage of ossification. In the first stage the skin and subcutaneous tissue may appear normal, but the underlying muscle is swollen and edematous. Microscopically, an extensive infiltration of the intra- or intermuscular connective tissue is seen. This infiltration is especially marked about the blood vessels. In the second stage the embryonic connective tissue becomes organized and forms adult connective tissue, which finally retracts and becomes a hard, fibrous swelling. The muscle fibers in this mass are scattered, more numerous at the periphery of the tumor, where microscopically many of them appear normal. Towards the center of the mass the fibers, when present, seem atrophied or swollen and frequently their cross striations are lost. The sarcolemma nuclei may be markedly increased; some may even resemble giant cells. Other cells are also seen which resemble cartilage cells and later produce hyalin cartilage. Osteoid trabecule, in which formative cells representing osteoblasts are found observed. New bone is also formed in the fascia, ligaments, tendons, and preexisting bones.

The chief histologic change shown in the case of Weber and Compton was the invasion of the striped muscle tissue and the normal connective tissue belonging to it by a newly formed fibro-cellular connective tissue which in its features resembled rather a fibromatous hyperplasia than a product of ordinary inflammation. The muscle fibers were undergoing degenerative changes, tending to split up longitudinally into fibrillæ, and in parts showed atrophy.

Treatment.—Medical treatment has been of no service. Hydrochloric acid, phosphoric and lactic acids have been employed, as well as mercury, potassium iodid, arsenic, and the salicylates. Thyroid extract has been of no avail. Fibrolysin has been tried, but with no beneficial results. In fact the tumor formation has occurred at the site of injection.

Surgical inteference has been very unsatisfactory, as there has been a recurrence at the site of removal in almost every instance.

The most important point as regards treatment is prophylaxis. If precuations against trauma and cold are taken, the affection may at times be prevented from spreading.

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ISCHEMIC MUSCULAR PARALYSIS AND MUSCLE CONTRACTURE

Ischemic muscular paralysis and muscle contracture, or Volkmann's contracture, is a deformity which was first described by Volkmann[1] in 1875. The condition was also described by Leser[2] in a monograph issued in 1884.

Volkmann's contracture is a condition which occurs generally in young children. It consists of a contraction of the fingers and sometimes of the wrists with partial loss of power in the muscle of the forearm coming on rapidly after a severe injury usually in the region of the elbow joint. The deformity is due to changes in the flexor muscles with injury to the peripheral nerves caused in many cases by tight bandages and pressure of splints.

Symptoms.—These come on rapidly, usually in half a day, or less in very severe cases, following the bandaging or splinting of the forearm or arm. The fingers frequently swell, and in some instances become blackish in color in a relatively short time. Although many authors claim that there is severe pain which may affect the whole arm, Dudgeon[3] states that pain is rarely present. In fact the absence of this symptom may be the cause of the splints not being loosened more rapidly. Frequently there is present some tingling of the fingers and a feeling as if the hand had fallen asleep. The course of the acute stage is very rapid so that the condition often reaches its height within a few hours. On removal of the splints the forearm is found tender, sometimes extremely so. At times sloughing of the skin may be present. When the dressing has been removed, sometimes 24 hours or more after it has been applied, a secondary reaction frequently occurs with an increase in the swelling of the musculature which may reach its greatest height in 24 to 48 hours. After this it gradually disappears, leaving a characteristic deformity. The fingers, which are at first simply flexed, soon pass into the characteristic position. With the wrist extended, the metacarpal phalangeal joints are extended likewise. The interphalangeal joints of the fingers and the terminal joint of the thumb are strongly flexed, so that the tips of the fingers touch the lower part of the palm, and no reasonable amount of force seems capable of straightening them. As soon as the wrist joint is flexed to a right angle, the interphalangeal joints can be easily extended. In very bad cases the wrists become strongly flexed so that they are incapable of extension. The hand is pronated, and the forearm is generally semi-flexed. The flexor muscles of the forearms seem hard and firm and become wasted. A thickening is sometimes felt in the soft tissues, usually in relation to the pressure sore which may be present. There may be either normal sensation or partial or complete anesthesia.

Dudgeon describes three groups of the deformity: (1) Pressure contraction of the fingers. (2) The hand in the typical position. (3) Strong flexion of the wrists and fingers without any power of extension. There are no true trophic lesions in pure ischemic paralysis. The hands are frequently cold and blue and the skin smooth. Often when the arm is allowed to hang down, the hand becomes swollen. In well-marked cases there has been observed some shortening of the bones which has been explained by Turney on the basis that the growth of bone proceeds only *pari passu* with the amount of support it has to give to its attached muscles. The disturbance of function varies. In the severe cases most of the muscles of the forearms and hands, flexors, extensors, small hand muscles as well as the pronators and supinators are involved, so that the hand cannot be used at all. In less severe cases, which are relatively more frequent, flexion only is interfered with, while in still lighter cases practically all of the muscles may be used to some extent.

The electrical reactions of both muscles and nerves are normal but may be changed, in that in very severe cases there may be no response to electric stimulation.

The cause of the condition is probably a combination of pressure fixation and ischemia, and is the result of prolonged fixation of forearm fractures by any kind of apparatus that interrupts the free circulation of blood through the muscles and nerves of the part. In some cases it is a combination of splint pressure and a large effusion into the soft parts. The sloughing or other signs of pressure sore is merely an indication of the amount of pressure the parts have been subjected to. It is not, as Davies-Colley[4] thought, the cause of the contracture.

Volkmann considered the condition as depending upon the diminution of arterial blood to the muscles occurring simultaneously with venous congestion, which hastens the paralysis. The muscle fibers deprived of oxygen perish, and the contractile tissue coagulates, breaks up, and disappears. The degree of paralysis depends upon the length of time the circulation has been arrested.

The pathology of the disorder is that of a primary necrosis of the muscle with secondary changes in the interstitial tissue which follow the destruction of the muscle and tend to contracture. The diagnosis is usually easy. The history of trauma with subsequent application of the splint or cast and the onset of paralysis, all occurring at the same time, the normal electrical reaction and sensation, and the characteristic deformity proclaim Volkmann's contracture. Ulnar, median, or musculo-spiral nerve paralysis must be differentiated from this condition. The contractures following anterior poliomyelitis and Little's disease resemble it only distantly.

The prognosis, according to Volkmann, is bad, especially in cases where the hand and fingers are implicated. Bruns points out the fact that considering the pathology it is not surprising that the severe cases offer a poor prognosis. In the milder cases, however, good results are obtained. Anderson gives a prognosis absolutely contradictory to Volkmann, and Dudgeon bears him out in the statement that the lesion is seldom of a permanent character.

Treatment.—The treatment consists of massage, electricity, passive movements, and baths. Of all these methods, methodical massage persistently given over a long period of time, for a number of months, if necessary, is the best. Surgical treatment has in many cases given excellent results.

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TUMORS OF THE NERVES

Nearly a century ago Odier[1] recognized the fact that tumors may grow from nerves. Tumors arising from the peripheral nerves or their vicinity are not common, but compared to their frequency in adults are relatively frequent in children. Tumors of the nerves, or neuromata, have been classified according to Virchow into true and false neuromata, according as the nervous tissue (medulated and non-medulated fibers) plays an essential part in the formation of the tumor or not. True neuromata are tumors arising from or composed of actual nervous tissue and the discovery of ganglion cells in one group of these tumors (Knauts,[2] Beneke,[3] and Busse[4]) confirms the existence of these rare growths. Knauts ascribes these tumors to the sympathetic nervous system. Bruns[5] states that these tumors may be single or multiple and that of the multiple true neuromata two cases had been reported in children, one by Knauts and one by Kredel[6] and him. Solitary true neuromata have been reported by Kredel and others.

The false neuromata are tumors which have their origin in the peri-, epi-, or endoneurium. They may be neurofibroma, neurosarcoma, neuromyxoma, etc. The false neuromata develop upon all the nerves but chiefly upon the spinal. They may be limited to the area of the nerves, as to the brachial plexus, the cauda equina, or may be distributed over the nerves of the whole body. Several forms of multiple false neuromata are found. Among them are included Ranken or plexiform neuromata, neurofibromatosis universalis or Recklinghausen's disease and elephantiasis neurosa.

True neuromata are exceedingly rare and are found practically only in connection with the sympathetic system, in the ganglia, within the thoracic or abdominal cavities, or the small ganglia of the peripheral sympathetic system along the blood-vessels and lymphatics. They contain, as a rule, ganglion cells and non-myelinated nerve fibers. They are benign in character but may infiltrate the surrounding tissue. They vary in size from that of a pea to a child's head and in consistency may be relatively soft or very hard.

The multiple true neuromata have been chiefly observed in children. They have been distributed in the peripheral part of the body, under the skin and in the subcutaneous fatty tissue or in the muscles. They are very numerous and in Bruns' case 160 were counted. The hands and feet only in these cases were unaffected. They were unattached to the skin or the underlying tissue and when pulled upon gave rise to no pain. The children otherwise were quite well. These tumors grow very slowly and the nervous system apart from them is quite normal.

Solitary true neuromata are never found in the periphery but only in connection with the sympathetic ganglia in the chest or abdominal cavities.

False neuromata develop from the connective tissues of the nerves. Their etiology is obscure. They generally occur in the first half of life and in many instances have been hereditary. Solitary neuromata may be found on any of the spinal or cranial nerves. Generally they are fibromata and of a benign character. Occasionally they may become myxomatous or cystic. The smaller tumors are generally spindle-shaped and elongated in the axis of the nerve from which they grow.

The most prominent symptom is pain which radiates from the tumor to the periphery of the nerve. It is generally intermittent, but may become continuous and agonizing in character. Movements of the limb may bring on the pain and the tumor is tender to palpation. Paresthesia is occasionally observed, but other changes in sensation and variation of reflexes is rarely present. It is characteristic of the tumors that they are movable in the transverse but not longitudinal axis of the nerve. The location of these tumors upon special nerves, as upon the optic or other cranial nerves, may give rise to further neurologic symptoms, as may be seen in the neurofibromata of the eighth nerve. When the tumor originates from the epineurium it usually lies to the side of the nerve, but when its origin is the endoneurium it penetrates the nerve and splits it into secondary bundles. The structure of the nerve fibers passing through the tumor is as a rule unaltered, but the myelin sheaths may show some changes.

The only satisfactory treatment is the removal of the tumors which produce symptoms. At times this may be done without interference with the nerve upon which the tumor may be placed, but often it is necessary to resect the nerve and to perform nerve anastomosis.

Multiple neuromata, or general neurofibromatosis is a condition in which large numbers of tumors may be found on many or all of the peripheral nerves.

Smith[7] has reported a case in which 450 were counted on one limb and 2000 were present over the body. These tumors vary in size from a bead to a child's head. The origin of these tumors is usually from the endoneurium. They are of spherical, elliptical or nodular shape. Histologically they are made up of connective tissue elements as are the solitary tumors and may be soft or hard. They grow slowly. In many instances they are hereditary, and in a still larger number of cases the congenital origin of the condition has been established.

All of the varieties alluded to above, plexiform neuromata, pachydermatocele, or neuromata elephantiasis, and multiple neuromata neurofibromata or molluscum fibrosum (Recklinghausen's disease), as well as tubercula dolorosa, are all expressions of the same pathologic tendency.

Neuromata plexiforme and pachydermatocele are clinical curiosities. In the former there is a diffuse hypertrophy of the connective tissue of all the branches of a nerve and the whole being imbedded in fat or in loose connective tissue gives rise to an appearance of a tumor. In the latter, not only are the nerves involved, but the subcutaneous tissues and skin likewise are thickened, and at times even the bone is involved.

Recklinghausen's disease is apparently congenital, occurring mostly in dwarfish people. Sometimes they have impaired intellect. It is characterized clinically by numerous cutaneous nodules which are sessile and painless. Frequently in addition to the skin phenomena there are neuromata of other nerves as well.

Neurofibromata have occurred on all the cerebrospinal nerves and sympathetic nerves except the optic and olfactory, which are essentially outgrowths of the brain. The fifth and eighth cranial nerves are those most frequently involved within the skull, but when we include all the peripheral nerves the order of frequency seems to be: Vagus, abdominal, sympathetic, sciatic. The intraspinal tumors usually arise from the roots, especially the posterior ones, and when the cord is invaded it is usually in the region of the entry of the posterior roots.

It has been clinically ascertained that neurofibromatosis is frequently associated with more or less mental impairment, imbecility and somatic stigmata of degeneration.

Several types of foci of disease are encountered in the cerebral cortex of many of the cases of neurofibromatosis. Bassoe[8] described areas with hyalin proliferated vessel walls and perivascular fibrous tissue and, secondly, minute areas containing polymorphous, rather large glia cells in heaps. The latter areas have given rise to much controversy since Bielschowsky[9] and others have suggested that they are identical with the lesion characteristic of the tuberous sclerosis of Bourneville. Bielschowsky describes the tumors of multiple neurofibrotosis as resulting from a similar blastomatous proliferation of the cells of the sheaths of Schwann. Nieuwenhuijse[10] does not see any striking similarity between the brain changes in neurofibromatosis and those of tuberous sclerosis, and he points out that the cutaneous tumors in the two diseases are different in type. He arrives at the conclusion that the two diseases are entirely separate.

Of special interest is the conclusion of Verocay[11] who thinks that the tissue of neurofibromata is not true connective tissue, but a neurogenic embryonal tissue the specific cells of which are capable of producing ganglion cells, glia and nerve fibers; hence, the occasional occurrence of ganglion cells in neurofibromata and the observed association with glioma and syringomyelia may be explained.

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CHAPTER CLXXXII

THE LEPTOMENINGITIDES

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Anatomy.—Childhood is the period of life which is most susceptible to the development of both the tuberculous and the suppurative types of meningitis. The infection becomes diffused through the tissues which enclose the cerebrospinal fluid. A brief outline of these structures will be of help in the later discussion of the pathologic changes.

The arachnoid membrane lies beneath the dura and subdural space. It is a thin fibrous structure doubly lined with epithelium, forming the external covering of the subarachnoid spaces, and furnishing sheaths for the nerve fibers. There are few blood-vessels and no nerve tissue in the arachnoid. It is intimately connected with the underlying pia mater over certain regions of the brain convexity and hemispheres, but over the posterior part of the brain base the arachnoid membrane helps to form large subarachnoid spaces, and these with the small spaces formed by the loose connective subarachnoid tissue, contain much of the cerebrospinal fluid.

The cisterna magna is the subarachnoid space between the cerebellum and the medulla, anteriorly communicating through the foramen of Magendie with the fourth ventricle. Below is the posterior arachnoid space of the cord.

The other large basal subarachnoid space is in front between the medulla, pons, interpeduncular space, crura cerebri and the optic chiasm and temporal lobes. This large space communicates directly with the anterior subarachnoid space of the cord, above it is in communication with numerous small spaces of the optic chiasm, Sylvian fossæ, corpus callosum, and behind the cisterna magna.

The pia mater covers the outer surface of the cerebrum and cerebellum with its thin, highly vascular membrane composed of delicate fibrous connective tissue, surrounding and accompanying the vessels as they enter the brain tissue. It follows the depressions of the brain fissures and forms the choroid plexuses and velum interpositum. Branches of the cranial nerves are found in the pia accompanying the vessels. Perivascular spaces or sheaths communicate with the subarachnoid spaces. The pia is therefore the inner membrane of the subarachnoid spaces.

The superior choroidal tela (velum interposita) and the inferior choroidal tela are processes of the pia communicating with or forming a membranous covering for the ventricle and masses of convoluted vascular choroid plexuses.

The fluid circulates through the ventricles, cisterns, subarachnoid spaces, bounded by the membranous walls, is emptied into the venous sinuses of the head or into the sheaths of the cranial nerves. Infection of these areas is always of grave import.

TUBERCULOUS MENINGITIS

Pathology.—Of all the forms of miliary tuberculosis, involvement of the central nervous system produces the most symptoms. The lesions in other portions of the body, with the possible exception of tuberculous peritonitis, are frequently not recognized until autopsy. Fully one-half of the patients dying of tuberculous meningitis show an active focal process. In all cases lesions elsewhere in the body will usually be found at a complete autopsy.

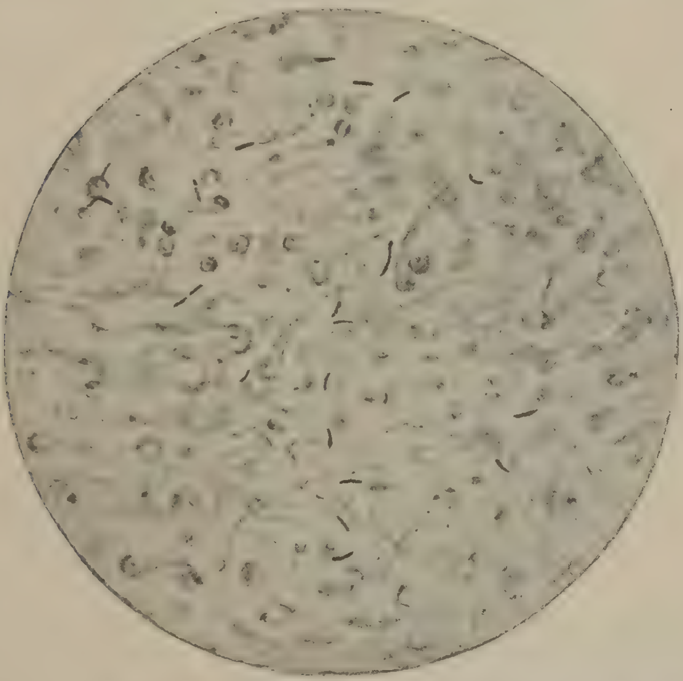


Fig. 64 *A*.—Tuberculous meningitis. Drawing from slide showing bacilli and broken-down lymphocytic cells from basal exudate in tuberculous meningitis.

Changes within the Cranium and Spine.—The physical changes due to tuberculous meningitis are always secondary to the spread or coexistence of a general miliary tuberculosis. Miliary tuberculosis of the central nervous system localizes itself in the pia mater and arachnoid of the brain and cord, in the cranial nerves, and by contiguity in the adjacent and dependent areas as hereafter mentioned. Tubercle bacilli may be found in the cerebrospinal fluid.

The gross changes in the brain are frequently so slight that they seem to be out of proportion to the severity of symptoms observed during life. But the microscopic examination reveals minute lesions which along with the general miliary tuberculosis easily explain the exitus of the child.

The area involved increases with the course of the meningitis, but at autopsy the brain base is found involved in all cases, the cortex in two-thirds and the choroid plexus in about one-third of the cases.

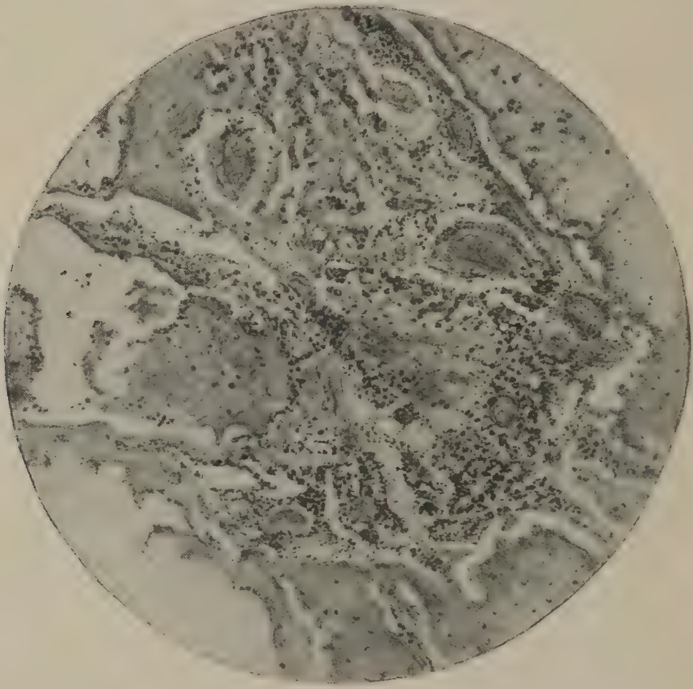


FIG. 64 *B*.—Tuberculous meningitis. Drawing made from a microscopic section of the pia-arachnoid tissues in the neighborhood of a blood vessel, showing a tubercle with some caseation and extensive round-cell perivascular infiltration of the small lymphocytic type; there are no giant cells in tuberculous meningitis in children.

The principal pathologic lesions are:

(*a*) Tubercle formation.

(*b*) Inflammatory gelatinous exudate.

(*c*) Development of granulations in the ventricular walls; a varying amount of internal hydrocephalus.

Grayish miliary tubercles are found at an early stage in the pia mater and in the arachnoid tissues, but they are often of microscopic size. They are arranged along the blood-vessels, especially in the depressions between the convolutions. The vascular distribution in the fossa of Sylvius and the cerebral and cerebellar base contain these tubercles in larger number than the convexity. The pia is involved in the exudative inflammation. It is congested, cloudy, bloody, edematous or may even be dry. Some of the tuberculous nodules undergo caseous degeneration and encapsulation.

The arachnoid space contains a sero-fibrinous, fibrino-purulent, seldom purely purulent exudate of grayish-yellow color and jelly-like consistency. The exudate as well as the tubercle formation is found chiefly at the cerebral base, in the arachnoid space and chiasm, in the cistern of the Sylvian fossa, and reaching to the peduncles. It sticks to the pia mater. It spreads along the vessels to the convexity, also by way of the connective tissue sheaths of the cranial nerves.

Miliary tubercles are also found in the walls of the ventricles and the choroid plexus. The ventricular fluid is increased and cloudy. The ventricles become dilated by accumulation of fluid which through pressure causes flattening and softening of the convolutions. More or less marked hydrocephalus may occur in tuberculous meningitis. The older writers thought that acute hydrocephalus was synonymous with tuberculous meningitis. Recently Blackfan has maintained that owing to the infrequent obstruction of the cerebrospinal canal marked hydrocephalus is not as common as was formerly supposed. The tuberculous process will be found microscopically to have spread from the pia to the cerebral tissue beneath it. There are evidences of encephalitis in the distention of the perivascular spaces, and in the perivascular round cell infiltration.[2] There are often capillary hemorrhages.

The above mentioned flattened convolutions are somewhat edematous, become roftened and infiltrated with tuberculous granulation tissue. They occasionally show punctate linear hemorrhages. The exudate contains small, large and degenerative lymphocytes. Large lymphocytes of the hyaline types will be seen in the venous walls. There will be found also desquamated endothelial cells resulting from the inflammatory proliferation in the arachnoid lining. The arterial walls and small veins are thickened, especially the endothelial lining of the vessels.

The tubercles which are found in the blood-vessels and arachnoid membrane consist of large nucleated cells, giant cells and lymphocytes in the periphery.

It has been found by Foot that meningeal and cerebral tubercles are formed experimentally in rabbits from cells which come from endothelium.[3]

Tubercle bacilli are found in the exudate and in the giant cells, the center of which cells may be necrotic as is also the center of the tubercle. Giant cells and typical large tubercle formations may be absent in early childhood. Small tubercles may be found in the brain substance. The tuberculous changes are found also along the blood-vessel walls in the spinal pia mater. There may be gross changes in the spinal arachnoid space due to thickening of the meninges, adhesions and occlusion or obliteration therefrom. There may be dilatation of the central canal of the cord.

A case of tuberculous meningitis is reported which presents a rare pathologic condition of the spinal meninges causing a dry spinal subarachnoid space. This was due to the diffuse infiltration of the pia arachnoid membrane, from the cervical to the lumbar region, with innumerable tuberculous granulations producing marked thickening of the membrane, and causing adhesions between it and the dura, almost completely obliterating the spinal subarachnoid space except in the spinal cul de sac. No case reports of a similar nature have been found in the literature reviewed.[4]

The cranial dura is usually involved only by being tense and bulging, but at times there will be small hemorrhagic or tuberculous formations on the inner surface. Likewise the inner surface of the skull may show some thickening.

Caries of the petrous portion of the temporal bone may be found. In generalized tuberculosis in children the autopsy frequently shows suppuration of the middle ear.

The cause of death if other than that of the general miliary tuberculosis is due to the destructive action of the tuberculous toxins upon the brain tissues, and to the greatly increased intracranial pressure which is incompatible with the maintenance of the blood supply, the nutrition and the metabolism of the brain.

Pathologic Changes in the Eye.—Choroid tubercles when present form part of the general miliary tuberculosis. Occasionally there are found retinal hemorrhages, also venous congestion of the eye-grounds and hyperemia of the papilli due to the existence of tubercles or to the internal hydrocephalus. A small percentage have choked disc from intracranial pressure, more frequently is present optic neuritis. Functional alterations will be mentioned under symptomatology.

Changes in the Spinal Fluid.—*Pressure.*—Through the introduction of Quincke's method of lumbar puncture, it was possible to study cases of tuberculous meningitis. It was found that the cerebrospinal pressure increased in the majority of cases ranging from 200 to 600 mm. (water). Exceptionally the pressure was lower or as high as 800 mm. depending upon the stage of the disease, much lower during the terminal stage. The pressure causes bulging of the fontanel and even a spreading of the sutures and flattening of the convolutions.

Appearance.—The clear colorless appearance of tuberculous cerebrospinal fluid differentiates it from that of other types of meningitis. This however is not always the case, as there may be opalescence or slight turbidity, or a yellowish or greenish tinge may be found due to xanthochromia in which case the fluid may contain much globulin and may coagulate immediately on withdrawal.[5]

Fibrin flakes are seen in some fluids. A fibrin coagulum—the so-called pellicle—is present in most fluids after standing undisturbed for several hours.

Cytologic and Chemical Changes.—The cell count is usually above normal, especially in the first portion; the greatest number of cells are found in the cases with more severe symptoms. The small mononuclear lymphocytes preponderate, may rise to 80 per cent. or more. Cells are few compared with other forms of meningitis, but in exceptional cases may run unusually high (1000 per cubic centimeter in one case of the author).

While it is generally accepted that an increase in the small mononuclear cells is characteristic for tuberculous meningitis, nevertheless an increase in the polymorphonuclear cells has been observed by some writers. In 154 children, reported by Dr. Josephine Neal, under two years of age, five showed 50 to 70 per cent. polymorphonuclear and two showed 70 to 96 per cent. In 151 children from two to ten years of age, three showed 50 to 70 per cent. polymorphonuclears, and two showed 70 to 95 per cent.

Globulin is usually present especially in the first fluid withdrawn.

The presence or absence of glucose as shown by the reduction of Fehling's is an important aid in the diagnosis. Very early in the disease the reduction may be normal, but as the disease progresses the reduction may become distinctly less or even absent. In poliomyelitis, encephalitis, or syphilis of the central nervous system, the reduction of Fehling's solution is usually about normal.

Microörganism.—The tubercle bacilli are specifically present in the fluid, more abundant in the late stage, though often difficult of demonstration. They will be found more often enclosed in the fibrin film.

Etiology.—Seasonal Incidence.—Tuberculous involvement of the brain is more common in the winter and spring months after which there is a marked decline. Acute respiratory disease is more common at this season, and its effect is to light up or exaggerate miliary tuberculous lesions which during such a phase may be carried to the meninges (pia-arachnoid). The seasonal influence may be produced solely through the lowered resistance which results from infectious respiratory diseases. There may also be a direct action upon the miliary processes by the changeable weather and its various influences upon the metabolism[6] of the body.

Influence of Age.—In the tuberculous population the ratio of cases with meningeal involvement is highest during childhood. In Germany the maximum incidence of deaths from tuberculous meningitis had been found in the age from one to two, but during the great war, this was advanced to the age of two to three years.[7] In tuberculous meningitis during childhood 60 per cent. will be under two years of age. Seldom will a case be found under three months.

As regards severity it is found that the younger the child the shorter the course, the less probability of remission and the fewer the signs of central irritation. That meningitis is the most frequent cause of death in the young tuberculous child is an accepted fact, though it must be recognized that it may be only one factor in the fatal issue in miliary tuberculosis.

During the years 1907 to 1909 at the St. Anne Children's Hospital, Vienna, there were autopsies on 109 children who died primarily of tuberculosis. Of this number 62 cases died of tuberculous leptomeningitis. This gives a percentage of 50.6 for leptomeningitis in deaths among children of all ages dying primarily from tuberculosis.[8]

Youth, then, is a predisposing factor in tuberculous meningitis, the explanation for which must be sought in the factors which make miliary tuberculosis the type which belongs especially to childhood.

Bovine Infection.—Experimentally tuberculous meningitis has been produced in dogs from tubercle bacilli virulent for guinea pigs and rabbits.[9]

In 30 cases of tuberculosis in children, reported by Gordon and Brown,[9] the cases of alimentary origin were bovine, those of pulmonary origin were human. Of these who died there were 18 in which the meninges were involved, three of which were bovine, 15 human in type.

In adults the percentage of bovine infection is small, slightly over 1 per cent. The percentage increases with the youth of the child, 25 per cent. in the ages from 5 to 15 years, 49 per cent. under five years of age.[10]

Exposure to Tuberculosis.—The cause of tuberculous meningitis is the spread of the infection with the tubercle bacillus to the susceptible cerebrospinal tissues. How the children are primarily exposed to tuberculosis is of great interest and importance. Infection from tuberculous milk is undoubtedly one source of the bovine type. The human type is to be sought for in exposure to diseased members of the family or household, who may have it in an active, latent or healed state by the time the child develops the disease. The mother as the nearest influence to the child will be first under suspicion. The search for tuberculosis must include any and all individuals who have lived in the household during the life of the child.

The writer has found active tuberculosis in the nursing mother, in the father, in uncles and aunts who have been living in the house, in servants, etc., so frequently that he feels that one is usually successful in finding the source. Statistics show that the mother is more commonly tuberculous, the father next in frequency, other relatives in a still smaller percentage. It is frequently found that some near relative has died of tuberculosis and in a small percentage other members of the family have died of tuberculous meningitis. In 355 cases in Escherich's and Moser's clinic 111 cases had been breast fed.

Children who are nursed by tuberculous mothers are particularly susceptible to tuberculous meningitis. According to the statistics of Herbert Koch, in children suffering from tuberculous meningitis under three years, 82 per cent. had been breastfed and 18 per cent. artificially fed.

Location of the Primary Focus and Method of Spreading.—The primary focus is found in the respiratory tract in nearly all cases,[11] less frequently in the mesenteric glands and in the bones.

Ghon concludes from autopsy findings in a large number of children that the primary tuberculous focus in at least 90 per cent. is in the lungs.

Miliary lesions are found at autopsy in the bronchial glands, lungs, spleen, liver, suprarenals, mesenteric glands, intestines, kidneys and bladder in the order named.[12] The average number of organs involved is from seven to nine.

From the wide dissemination of miliary tuberculosis, it is impossible to say from which focus the secondary meningitic infection comes. The channel of infection was thought to be a lymphatic one, coming from diseased glands or bones. It is now generally agreed that the route is hematogenous.

Influence of Trauma.—Nearly every history will contain the mention of a "fall" or trauma, which at the time was probably not regarded by the parents as being of consequence, but after the development of brain symptoms will be given due or undue prominence as the explanation for the child's illness. One should not disregard the information thus received though the causative relationship may not be capable of proof. A latent focus anywhere in the body may be so "broken up" by tissue injury that the bacilli are carried broadcast. More directly there may be a reduction of the resistance in the pia mater to the Koch bacillus after a fall or blow on the head. Either or both of these methods of dissemination may occur.[13]

It is now conceded that trauma can be considered a cause of spreading an existing tuberculosis to the meninges. This is considered the more probable if the interval has been between 3 and 12 days, and if the child before the traumatism had been known positively to have been in good health.

In four children cited by Barabas[14] the interval between trauma and death was from 16 to 23 days. When tissue is traumatized or embolus established it is easy for the tubercle bacillus to grow.

On the other hand, tuberculous meningitis does occur without known trauma just as a miliary process may be spread to any other portion of the body.

Trauma in other regions than the skull may be influential as a cause of dissemination. Operation upon tuberculous glands, operation or other traumata to tuberculous bones may free bacilli which are taken up by the injured blood-vessels.

A case of tuberculous meningitis which followed the production of artificial pneumothorax was reported by De Lille[15] as a warning against such traumatization in children in whom both lungs are affected.

Influence of Infectious Diseases.—Influences that reduce the resistance of the tuberculous child so that the disease is lighted up and a meningeal focus develops are the contagious diseases: Measles, pertussis, chicken-pox, diphtheria, scarlet fever; respiratory febrile diseases—pneumonia, influenza, etc. It is well known that the cutaneous tuberculin test is negative during measles. Whooping-cough as a forerunner of tuberculous meningitis is nearly as frequent as measles, and these two diseases stand in the first place. Tuberculous cervical nodes, and the other primary lymph gland tuberculosis may go on to death from miliary and meningeal involvement. Any exhausting febrile diseases may reduce the resistance and the reserve strength of the child to a degree that the spread of the miliary processes is stimulated.

Tuberculous meningitis followed grip in 80 cases seen by Morquio,[16] verified by the finding of tubercle bacilli in the spinal fluid in 80 per cent.

Congenital lues can be regarded as one of the predisposing causes of meningitis as it is of tuberculosis in general.

Two observers who have seen 20 cases of tuberculous meningitis in syphilitic children believe that lues in the young makes a ready soil for the tuberculous localization in the brain, and when both diseases are proven there should be active specific treatment.[17]

Symptomatology, Course and Duration.—Typical Cases. *Early Symptoms.*—The first change is seen in the character of the temperament and disposition. The child who may have appeared in the best of health becomes irritable, cries without cause, is easily frightened; because of fatigue cares nothing for playing, has loss of appetite, becomes constipated; has headache, disturbed sleep and dreams. This is succeeded by quiet and stupor, the anorexia and nausea so pronounced that it is impossible to get food taken or retained. Early vomiting is often mistaken for dietary indiscretions. Vomiting, constipation and other gastro-intestinal disturbances may resemble cyclic vomiting, typhoid fever or ileocolitis. Constipation becomes more marked or in the young infant there may be loose undigested stools.

There may develop pain in the chest, abdomen, extremities; and headache located in the vertex is discernible in older children. The child does not wish to be disturbed, light is annoying and photophobia develops, slamming the door or other noises are irritating, and there may be peculiar disturbances of the taste and smell.

Later Symptoms.—Of great significance are signs of motor irritability which now appear whereby the child grinds the teeth, sighs, groans, yawns, moves the lips and corners of the mouth, or utters the short hydrocephalic cry; blinks the eyes, wrinkles the forehead; has meaningless movements in grasping at nothing or at the bed clothes, picking the lips or genitals. Over the body will be found convulsive movements in evidence of a general hypertonus. Ankle clonus, a positive Babinski and Kernig sign, and resistance to all attempts at passive motion are usually present. Kernig's sign is said to be present in any meningitis in which the effusion is scanty. By this time stiff neck appears, a symptom which is predominant in tuberculous meningitis and which is accompanied by retraction of the head. In the children beyond the age of fontanel closing, the cracked-pot percussion sign of MacEwen will be positive and of value. The abdomen is retracted into the scaphoid or boat-shaped appearance, though in infants there may be a marked distention.

Convulsions of the grand mal type, petit mal seizures, localized convulsions of muscle groups are present in individual cases.

One of the most suggestive symptoms which now appears is strabismus; even before this symptom is pronounced, it may at times be produced while the head is flexed anteriorly, when there will be seen a transient unilateral or bilateral internal strabismus.[18] There is also a definite contraction of the pupils during the time that the head is flexed forward (Squires),[19] and a dilatation of the pupils when the head is extended backward.

There are frequently sudden appearances of areas of redness of the skin, and dermagraphia (cerebral tâche) is easily elicited. The child is hypersensitive, delirious or in mild coma.

The pulse is irregular with a tendency to become retarded more and more to as low as 50 per minute in spite of temperature increase. Slowness, irregularity and inequality are features of the pulse in tuberculous meningitis. The temperature may in a few cases be at or near normal, but usually there is an elevation of from one to three degrees, becoming gradually higher as the disease advances. A number of cases have been reported in which the axillary temperature was higher than the rectal, and due probably to bulbar involvement.

The respiratory rhythm is disturbed by the meningitic involvement, though there may be intrinsic influences in the diseased lungs. The marked feature is irregularity. This is noted late in the second as well as early in the third stage of tuberculous meningitis. Suddenly the respiration is quickened, then ceases for a few seconds, so that the total respirations in a minute do not exceed 12 to 16. This was described by Bouchut in 1862, and is of as great significance as are the pulse changes in this disease. Out of this type of breathing may be developed the Cheyne-Stokes respiration later in the disease. Biot's respiration is not so frequent in tuberculous meningitis as in other forms. It consists of periods of apnea, constant differences in force and rhythm, and of deep sighing respiration.

Attention has been called to the stimulation, inhibition or paralysis of the respiratory center from involvement in the meningeal disease. Intracranial pressure disturbs the rhythm, the respiration becoming wave-like due to diaphragm involvement, and later there follows Cheyne-Stokes breathing which is here a functional disturbance of cerebral origin.[20]

Paralytic Symptoms.—In the course of the disease there now will be found various paralyses, transient or permanent, especially in the cranial nerves, affecting most frequently the eye and facial muscles, causing strabismus and ptosis and facial palsy. Choked disc is found.

Hemiplegia sometimes develops, as also paraplegia; contractures and spastic paralysis may result. There is loss of control of the sphincters of the bladder and rectum.

Cerebral irritability diminishes; stupor increases, delirium disappears. There is increase in the intracranial tension as shown by the tenseness of the fontanel and by the spinal pressure.

Terminal Symptoms.—In the cases running the usual length of time there has developed a marked wasting of the body. The child lies in a deep coma, with abolition of all conscious movements and the reflexes, the pupils dilated, the arms and legs spastic, few if any convulsions, the temperature irregular, the pulse at the last changes, becomes fast, the respiration superficial and of the Cheyne-Stokes type and finally stops.

The duration is shorter than generally considered, the average being 17 days, the time being shorter in the younger child.

Atypical Cases.—In infants especially the disease may run its course rapidly and many of the symptoms elicited in older children cannot be obtained. It may develop in the course of an infectious disease or during the manifest presence of tuberculosis of the lungs, peritoneum, glands, bones, etc., when its presence will be masked by the illness or disability from which the child has been suffering.

The writer saw one case in which the disease was ushered in suddenly by the appearance of hemiplegia which disappeared shortly during the course of the typical early symptoms, and which might be confused with poliomyelitis. This is explained by the primary meningeal involvement of the motor area. Unilateral convulsions and aphasia may occur in these early suddenly appearing cases.

Chronic and remittent cases are reported in late childhood and adult life. Here the disease is probably localized as a meningitis en placques or as a solitary tubercle. According to recent statistics in this country solitary tuberculoma is rare in childhood.

Individual cases may have early eye palsy, headaches, or spinal symptoms, or the psychic manifestations may be exaggerated.

It has been suggested by Gilbert[21] that there can be a light form of tuberculous meningitis secondary to tuberculous choroiditis. In such cases there is general headache and stiff neck. The meningeal symptoms disappear in a few weeks.

Diagnosis.—**Spinal Puncture.**—The specific diagnosis depends for its final confirmation upon the finding of tubercle bacilli in the cerebrospinal fluid. Spinal puncture has now become a routine in the examination of all cases of suspected meningitis. The technique of this need not be described here. Whether the child is sitting, held over the shoulder of the assistant, or kept reclining on one side is to be determined by the facility with which the child can best be held quiet and the skill of the operator best obtained.

In rare instances dry tap results from the puncture and may be due to obliteration of the spinal lumen from local tuberculous disease above the point of puncture.

Skill should be acquired in its performance, especially as the spinal canal of young infants is more difficult to enter because of its small caliber. There is no contra-indication to the repetition of the puncture daily or on alternate days if from the first day's fluid no conclusions can be drawn, and if the procedure gives therapeutic benefit by lessening the symptoms of cerebral excitability.

Examination of Fluid for Bacilli.—The very careful technique of Hemenway[22] is recommended for the bacteriologic examination of the fluid by which tubercle bacilli are nearly always found.

The last of a liberal amount of fluid (60 to 100 c.c.) is kept unshaken in a tube placed in the incubator over night, after which the fibrin coagulin is removed by a platinum loop, decolorized and counterstained. If the film does not form, the sides of the test tube are scraped or the fluid is centrifuged. The average time consumed in finding the organisms by this method is one hour. As the disease advances the bacilli will be found in practically all positive cases.

Inoculation of a laboratory animal with the suspected fluid requires three or four weeks for confirmation and therefore is impractical from the clinical standpoint.

The isolation of the microörganism is obtained by specific stains, and the division into the two types of bovine and human is done by cultural and inoculation methods. A subject which has been of speculative interest is whether there is a change in the morphology of the strain of the tubercle bacillus found specific for meningitis. Novich[23] concluded from a large number of specimens that the tubercle bacillus found in spinal fluid does not have the fine beaded appearance, the polar granules, and the slender form which are characteristic of the bacilli coming from lesions in other tissues of the body.

Confirmation Tests in Spinal Fluid.—The fluid is clear or slightly opalescent, or may have fine flecks in it. The type of cells is predominantly mononuclear, the total cells are increased but not to any characteristic degree. Chemical and cytologic examination of the fluid is not specific for tuberculous meningitis.

The Noguchi globulin reaction is positive in the fluid of all meningitis cases. Pandy's reaction for globulin has recently proved valuable for determining abnormal amounts of albumin in any meningitic fluid and is obtainable before other evidences of meningitis are present. It is useful also because of its simplicity:

One drop of cerebrospinal fluid when added to 1 c.c. of 1:15 carbolic solution gives when positive a bluish-white cloud at the juncture of the two.

There is a typical zone reaction in the colloidal gold test in tuberculous meningitis of some value early in the disease.[24]

The complement fixation test will be positive in the spinal fluid and will be of value if reliably performed, especially where the tubercle bacilli are difficult to find.

A specific test for tuberculous meningitis deals with the appearance of an increased number of red cells in the fluid after an injection of 0.01 to 0.002 mg. of old tuberculin into the spinal canal.[25] One must in such a procedure exclude the possibility of traumatic hemorrhage.

It has been reported that in tuberculous meningitis xanthochromia may be accompanied by a few red cells in the spinal fluid.

Cerebrospinal Fluid Pressure.—Usually there is some increase in the spinal fluid pressure at least at the first puncture in the earlier stages of the disease, during which time the open bulging fontanel offers an easily elicited confirmation. The pressure readings have been discussed previously under the heading of pathologic changes in the spinal fluid. They give nothing distinctive for tuberculous meningitis.

Ventricular Puncture.—In the infant with open fontanel the diagnostic puncture of the ventricle is coming more and more into use, the removal of fluid from which serves for laboratory study as well as reduces temporarily the increased intracranial pressure.

Conclusion.—One must conclude that the examination of the spinal fluid offers no great help beyond the finding of the specific organism and a clear fluid, and that for practical purposes the history, clinical symptoms and course of the disease must for the present be relied upon for the ante-mortem diagnosis of tuberculous meningitis.

The Blood.—There is usually a definite leukocytosis in tuberculous meningitis. Counts of 10,000 to 20,000 are usually obtained though individual cases may run twice as high. The number usually increases with the progress of the disease. There is an absence of eosinophiles. The differential count shows that the existing leukocytosis is definitely due to increase in the polymorphonuclear cells, which are present in marked preponderance.

The red cells and hemoglobin decrease with the development of secondary anemia and emaciation.

The Urine.—This is decreased in amount, the specific gravity increased, contains some albumen, acetone and diacetic acid, and in case the kidneys are involved in the miliary processes, tubercle bacilli may be found. The general findings are not different from those of other severe infections or of miliary tuberculosis itself.

Cutaneous Tuberculin Tests.—The early opinion that the von Pirquet test is negative in all miliary tuberculosis is not borne out by later experience. Either a von Pirquet or intracutaneous reaction will be obtained in most of the cases if proper technique, repetition on various portions of the body and suitable tuberculin are used before the last stage of the disease is reached. The result of such a test does not of itself prove or disprove the presence of tuberculous meningitis.

Resumé of Principal Diagnostic Signs

1. Rigidity: Stiff neck, head retraction, spasticity and contractures of the extremities.
2. Eye symptoms: Delayed reaction to light, inequality of pupils, later a dilatation and absence of pupillary reaction, strabismus.
3. Positive Kernig sign. Knee-jerks are inconclusive.
4. Positive Brudzinski sign (drawing up of knees when head is flexed forward).
5. Binda's shoulder sign positive. (The chin is brought toward one shoulder, causing the opposite shoulder to move upward, forward and remain so placed until the head is released.)
6. Cerebral irritability.
7. Increased intracranial pressure and bulging fontanel.

8. Convulsive movements and paralyses.

9. Slowing and irregularity of pulse, rapid-pauseful disturbance of respiratory rhythm.

10. Bacilli (Koch) in cerebrospinal fluid.

In addition to these the evidence of tuberculosis elsewhere (the *x*-ray may give information of tuberculous processes in the lungs, mesenteric tuberculosis may have been proven on operation), the fibrin-forming clear spinal fluid with increase in cell count and the rapid and fatal course, give valuable evidence, while the autopsy findings will be accepted as final proof of the tuberculous variety of meningitis.

Complications.—Complications with purulent meningitis are rare in childhood. When the exudate in tuberculous meningitis is purulent it may be due to accidental mixed infection. Meningococcus infection may co-exist when there is an epidemic of meningococcic meningitis. Sabrazes[27] reported 350 cases in France among adults. Only 21 cases were collected by Faber in which purulent meningitis was a complication. His own case was the only one of influenza meningitis—a complication of peculiar rarity when one recalls the frequent influenza complications in tuberculosis of the lungs.

Death from hemorrhage into a cerebral hemisphere has been reported in a case of tuberculous meningitis (Reuter).

Typhoid fever co-existed with tuberculous meningitis in a case reported by Cole.

Cammarata reports a case of a three-year-old boy having symptoms of multiple sclerosis for a period of nine months: Tremor of the right hand, later anesthesia of right leg, and the development of a tremulous, spastic gait, intention tremor of both hands, a few weeks later he developed tuberculous meningitis with tubercle bacilli in the spinal fluid, and then died. Whether he had both diseases or the meningeal infection was a focal one is not clear.

Differential Diagnosis.—(1) From a localizing standpoint the diseases which must first be mentioned are those which involve the leptomeninges and the brain base.

In acute serous leptomeningitis and the meningism of infections such as intestinal intoxication and infections, typhoid fever, pyelitis, pneumonia, etc., the symptoms are explained by the prevailing disease, there is the absence of tubercle bacilli in the spinal fluid, the course is not characteristic of tuberculous meningitis nor is the outcome uniformly fatal.

Most of symptoms of tuberculous meningitis may be found in typhoid fever in children. The spinal fluid in both is clear. However, vomiting and convulsions, and scaphoid abdomen are rare in typhoid, while optic neuritis, choroid tubercles and a leukocytosis are present in tuberculous meningitis.

Acute purulent leptomeningitis and epidemic purulent leptomeningitis are characterized by cloudy spinal fluid and their causative pus-producing organisms.

Basal brain tumors and basilar syphilitic meningitis come into question and are diagnosed by their pathognomonic signs, pneumoventricular inflation, serology, longer course, etc.

2. Brain abscess follows suppurative otitis media, sinus thrombosis, compound skull fractures, etc.

3. The presence in recent years of what is now known as epidemic encephalitis has added another not rare disease which must be differentiated, and the task is a difficult one unless tubercle bacilli in the spinal fluid and evidences of miliary tuberculosis are obtained. The spinal fluid in encephalitis usually contains more sugar-reducing substances, the cell count is lower and the gold reaction is in lower dilution. Rarely the pia is involved in the perivascular infiltration, in which case the brain symptoms for a time may be similar.

Tuberculous meningitis shares with meningococcus meningitis the first position in point of frequency. The two must therefore be at once differentiated from the standpoint of the prompt therapy needed in the latter. For the clinical differences in the two the reader is referred to the description of the two diseases. For practical purposes the bacteriology and cytology of the spinal fluid is the deciding point. Worthy of note is the difference in the urinary output in these diseases: An oliguria occurs in tuberculous meningitis, a polyuria in meningococcus meningitis.

Prognosis.—There have been no proven cases of recovery in the first two years of life. These comprise about 60 per cent. of all cases. In later childhood reported recoveries have a suspicion attached to the specificity of the diagnosis. Practically all cases of reported cure are in adults in which the recovery is sometimes claimed to be complete, others finally died of tuberculosis in other portions of the body. When one reads the history of a reported cure, he finds that the meningitis has not been of the basilar type and with the evidences of cerebral paralysis. No doubt there are instances of tuberculosis of certain portions of the body which may have an intercurrent meningismus of a toxic or serous origin. Many years ago Barth[28] had the opportunity of examining the brain 25 years after a healed tuberculosis of the meninges—in which he found fibrous changes in the pia and old nodules along the blood-vessels which were evidences of a former meningitis. There are numerous writers who report cures, but it must be required for proof that tubercle bacilli be found in the cerebrospinal fluid, or that at autopsy the scars or fibrous tissue of an earlier tuberculosis are found in the pia arachnoid. Lesions in the meninges of tuberculous origin are probably not of the miliary type if remissions or recoveries occur. The general miliary tuberculosis makes the case hopeless.

Prophylaxis and Treatment.—Prophylaxis offers at present the only hope for the reduction of mortality in this disease; early removal of the child from the tuberculous environment; safeguarding the food by urging the nursing from healthy mothers; by the use of safe milk from a tuberculosis-free herd, and especially in bottle infants the boiling of all milk; protection of the child from measles, pertussis, influenza, pneumonia, and all other debilitating diseases which predispose and lower the resistance; the feeding of optimal amounts of good food; the maintenance of hygienic standards for abundance of fresh air and sunshine. The foregoing idealistic factors are essential for the reduction of the number of cases of this dreaded disease.

Tuberculous meningitis in the child is as yet without proven cure. Whatever benefit has been obtained has been through intraspinal or intraventricular puncture, either by the reduction of pressure, the removal of bacteria-laden fluid, the injection of air intraspinously and into the ventricles[29] (for which there seems to be some reason in the analogy from the open treatment of tuberculous peritonitis), the injection of a non-specific serum (anti-meningococcic). These agents have been used, and again it is in adults that there has been any success. The autoserotherapy of tuberculous meningitis has been tried in a few cases. Cerebrospinal fluid was withdrawn and injected subcutaneously with apparently beneficial results in three children one of whom was said to have been cured.[30]

For the present one must be satisfied in relieving the pressure symptoms especially the headache, convulsions and paralysis by an occasional spinal puncture and by the relief of special symptoms as they arise. Sedatives, body hygiene and food should receive all possible attention.

There is left only to say that the most useful place for research in the whole field of tuberculosis in childhood is found in the problem of a successful treatment for tuberculous meningitis. Bone, joint, peritoneal and glandular tuberculosis is curable. General miliary disease—the essential tuberculosis of childhood—is rapidly fatal. Whatever chance may ever be found in miliary tuberculosis will be through the promptness of the treatment with a hitherto-undiscovered specific.

PURULENT MENINGITIS (EXCLUSIVE OF MENINGOCOCCUS INFECTION)

There will be considered under this heading only those types of infection in which the leptomeninges are invaded by pyogenic organisms; serous meningitis is treated in another chapter, as is also epidemic (meningococcus) meningitis.

Pathologic Changes.—**Gross Appearances.**—The offending organism localizes in the pia-arachnoid system. Hyperemia and serous exudate soon result. When the brain is exposed at autopsy a striking alteration from the normal brain surface is presented, especially at the base. There is seen a diffuse cloudy or yellowish exudate which is fibrono-purulent, brownish sanguino-purulent or strictly purulent of a creamy or greenish color. This is found loosely adherent to the surface of the membranes, most marked at the base of the brain, and continuing downward into the anterior subarachnoid space of the cord. It may also exist as free purulent fluid mixed with the cerebrospinal fluid in the subarachnoid spaces. It consist of pus, fibrin and serum. The purulent exudate may extend outward and upward where it will be seen as streaks of pus in the furrows of the brain convexity and along the wall of the vessels in the venous depressions. As the suppurative process extends, the convolutions are covered with the thick sticky purulent material, and the increased intracranial pressure flattens the convolutions.

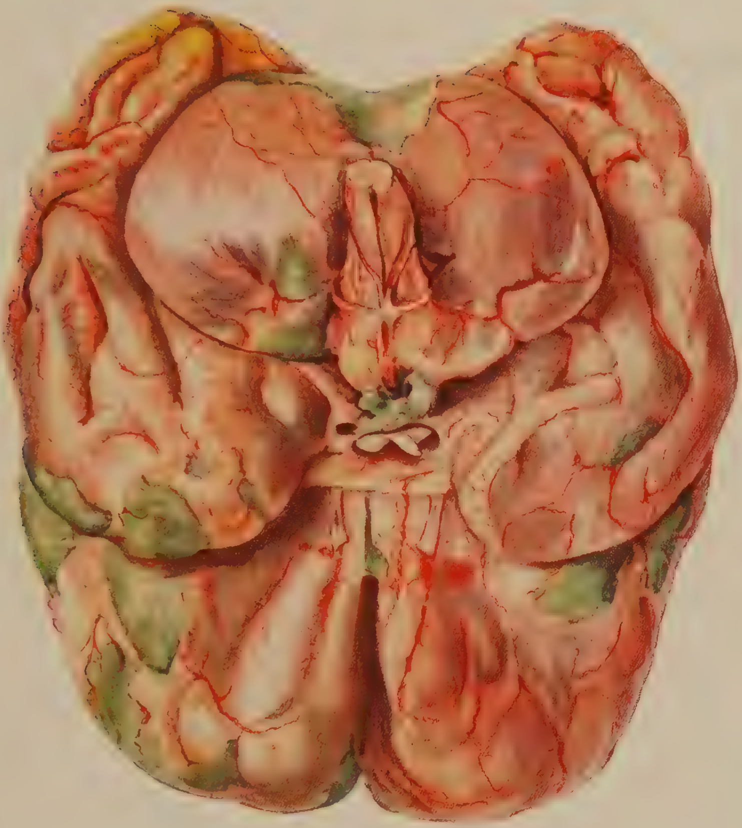
Minute Appearances.—The exudate is made up of serum and polymorphonuclear cells infiltrating and surrounding the blood-vessel walls; red cells and fibrin if hemorrhage has occurred; endothelial cells showing phagocytosis may be present; the connective tissue of the arachnoid membrane is in like manner infiltrated and its fibers proliferated. The fibrous bands show a proliferation of their endothelial lining, which becomes in some places necrotic. There may be edema of the membranes. The underlying brain substance becomes involved in the inflammatory and degenerative processes by apposition or by the vascular and pial prolongations entering the cortex, and may have many small hemorrhages. Under the pia and in the adjacent brain substance of the cortex will be found exudate which travels along the blood-vessel sheaths; cellular infiltration, edema or pulpification of the superficial brain tissue will be seen. The small veins become thrombotic, and the cranial nerve fibers and ganglionic cells are invaded by the purulent exudate and become necrotic. If the fibrin of the exudate becomes organized it may block escape at the foramen of Magendi and cause internal hydrocephalus. The end result is the production of a purulent leptomeningo-encephalitis.

Etiology.—Acute suppurative rhinitis and even otitis media is so often disregarded in many children that these causes may be overlooked, and the sudden appearance of meningitis seems to visit an apparently previous healthy child. A lymphatic or hematogenous route is undoubtedly the channel by which such infections reach the meninges. Disease of the tonsils, bronchitis or rhinitis, or an intestinal infection may be so insignificant as to be disregarded as the primary focus. The pharynx, nose or Eustachian tube may be the source of the infection.

In such manifest diseases as pneumonia, pleurisy, grip, influenza, typhoid, endocarditis, rheumatism, scarlet fever, the developing or secondary meningitis may be obscured for awhile by the primary disease. Bacteremic diseases and emboli carry the organism to the membranes of the brain.

Much more discernible and common in its causation is the type of purulent meningitis following the transmission of the infecting organisms from neighboring suppurative processes by extension. The most evident instance of this variety is the spreading from an otitis media. The nasal sinuses, cranial sinus thrombosis, abscess of the scalp, neck, brain, orbit, bones of skull, mastoid and dura may carry their suppurating processes directly to the pia.

PLATE I



Purulent meningitis. Appearance of the brain base in a one-year-old child dying of purulent meningitis of otogenous origin. The infant was brought to the hospital because of diarrhea, vomiting, and fever. Cerebral symptoms were mild and insidious, and at the first spinal puncture there were found only 4 or 5 cells per centimeter. Five days later, with the development of daily convulsions, deep stupor, slight stiff neck, and positive Kernig, a second spinal puncture was performed, and this showed a cloudy fluid containing 3700 cells, mostly polymorphonuclears; diplococci were found in the spinal fluid. Only a few drops of fluid were obtained at ventricular puncture, and at autopsy the ventricles were not dilated. Daily injections of antimeningococcic serum intraspinally, intramuscularly, and once intravenously were without effect.

The entire brain base as well as the convexity up to the longitudinal fissure contained islands of grayish-green sticky pus, and the sulci especially were filled with it. Both mastoids and middle ears were found to contain pus, but there was no evidence of direct extension from these through the dura. The mode of transmission seemed to be along the nerve sheaths or blood-vessels. The smear and cultures from the pus covering the brain showed the same Gram-positive extracellular diplococcus—a pneumococcus. Organisms were scarce in the spinal fluid. Diplobacillary forms were sometimes found. This child did not have pneumonia.

The extension of an erysipelatous infection of the scalp and face to the meninges though rare is always feared. Furuncles, osteophlebitis, infected scalp wounds, open skull fractures, basal fractures, post-operative mastoid or antrum suppuration are potential if not frequent causes for diffuse meningeal involvement.

Purulent meningitis is the most frequent form in early infancy. The infecting organisms in order of frequency are the pneumococcus, streptococcus, influenza bacillus, colon bacillus, staphylococcus pyogenes aureus, Friedlander diplo-bacillus, bacillus pyocyaneous, typhoid and paratyphoid bacillus, diphtheria bacillus, and rarely the gonococcus.

Meningeal infection is more common in childhood because of the greater susceptibility at that age of life. Seldom are infections of the meninges primary even then, as there are natural resisting barriers to infection of the leptomeninges. Intracranial complications after suppurative ear operations are seldom seen under the age of five years. The route in otogenous cases both before and after operation is probably by the labyrinth and dura.[31]

Symptoms.—The disease is characterized by sudden onset which will be seen in the early vomiting and loss of appetite, and the appearance of marked fever. The temperature usually remains high throughout the course. The pulse is rapid and regular, until changes are brought about by marked increase in the cerebrospinal pressure. The pulse may or may not increase with the temperature. The respiration is irregular and quality unequal, and in the well-advanced case the Biot type of respiration is frequently found. The reflexes are equal but disappear later in the course. Restlessness, delirium and later stupor, stiff neck and head retraction are usually present. There is headache or pain on raising the head. In young children convulsions of general or localized type are usual. The eyes show: Pupils dilated, fixed, irregular, loss of light reflex, conjunctivitis, strabismus, ptosis, muscle palsies. There is retention of urine.

Diagnosis.—The difficulty of making an early diagnosis of meningeal disease in childhood is recognized. Most of the symptoms of meningitis may be found at times in other infectious diseases. Convulsions at the beginning of infections, some of which are trivial in themselves, are common during childhood especially in the spasmophilic diathesis. The ease with which convulsions occur may be due to the rapid increase in cerebrospinal pressure resulting from hyperemia of the fluid-producing tissues.

Moderate increase in cell count is of no diagnostic import unless the bacteriology of the spinal fluid and the clinical course of the disease are confirmatory. Spinal puncture has made possible the differentiation of the numerous purulent types which can now be described under their various causes.

All investigators of experience will admit the difficulty which is encountered at times in performing lumbar puncture especially in infants whereby the accidental piercing of a blood-vessel brings only blood or a bloody spinal fluid. This is explained by the fact that in cases of small amount of fluid and low pressure the dura is pushed ahead of the needle against the wall of the canal.[32]

The Various Types of Purulent Meningitides.—It is advisable to discuss separately the different types of purulent meningitis not because there are many pathognomonic features which distinguish one variety of meningitis from another, but because the primary infection, the bacteriology and to a less extent the spinal fluid characteristics are necessary to an understanding of each one.

Influenzal Meningitis.—Influenzal meningitis is probably incorrectly named, as it does not depend upon the existence of a so-called influenzal epidemic for its appearance. It has been singularly absent during the pandemics of influenza, but occurs sporadically or at most during the late fall and early winter months, varying in different years, when so-called grip is common. During the epidemic of 1918, observations in New York City revealed only one case of influenzal meningitis. It can be said to be caused definitely by the bacillus of Pfeiffer, and while certain cases may be apparently primary it is usually preceded by pleurisy, pneumonia, otitis media, abscesses, osteo-arthritis, rhinitis, sinusitis, or a general blood infection. The disease is then rarely a primary, usually a secondary suppurative inflammation of the pial and arachnoid tissues.

Pathologic Changes.—A massive purulent meningitis is the rule, with the exudate usually covering the base or involving also the cortex, markedly over the frontal lobes, especially along the blood-vessels. There may be either microscopic or gross evidences of inflammation especially in the perivascular spaces of the brain substance, hemorrhages of large or small size may occur in the pia and in the spinal canal. There is a round cell infiltration of the vessels into the gray matter of the cord. The exudate is of a greenish-yellow color, markedly purulent, the characteristics of which are the large number of cells—polymorphonuclears, the mononuclears and large endothelial—many bacilli, small hemorrhages, and the little fibrin. The blood-vessels may be infiltrated and thrombotic and the exudate so organized as to cause an obstructive internal hydrocephalus if the patient lives long enough. The cranial dura is usually involved, and is congested. The cord contains less exudate than the brain.

Etiology.—There have been found to be numerous strains in the bacilli isolated from influenzal meningitis and these are usually different from those found in the respiratory tract (Rivers, Cohen, Fitzgerald, and others). Neal thinks that the organisms belong to the same strain. When pneumonia co-exists it is found usually in the posterior portions of the lower lobes.

The organism which is isolated is Pfeiffer's bacillus. It is Gram-negative, grows only on media containing hemoglobin, slender but showing constant tendency to varying size and shape, from small diplococci to short or long thready bacilli, staining distinctly at the poles, usually extracellular.

The route of infection is hematogenous.

Age is a predisposing factor; even the newborn are susceptible and most cases occur during the first two years of life. The disease is essentially one of infancy. Meningitis occurred in twins five days old, which indicates that the incubation period is somewhere within five days.[33]

Diagnosis.—The febrile course is sudden in onset and shares with influenza itself the frequent and quickly varying degrees from high to normal changes, but when pneumonia or purulent pleurisy co-exists the temperature will usually remain high. Likewise will there be variations in the respiratory rate dependent upon the degree of temperature elevation, the presence of pneumonia, and the involvement of the respiratory centers, when irregular, shallow, sighing, pauseful respirations may also be present. The pulse is fast and may be irregular.

Evidences of increased intracranial pressure, bulging fontanel, headache (especially if the convexity is involved), vomiting, restlessness, abolition of reflexes, convulsions, neck signs (if the disease involves the base) are found, a positive Kernig and Babinski sign frequently present.

The eye symptoms are photophobia, relative differences in the pupils, absence of reaction to light, nystagmus, and the eye-grounds showing a neuroretinitis.

Stupor, delirium and coma are pronounced as the disease advances.

The spinal fluid soon after withdrawal forms a thick clot of white or yellow sediment above which the fluid may be clear or turbid. The fluid is at first ground-glass in appearance and later in the disease is thick from its purulent content. The indole test is an aid to the rapid diagnosis of the spinal fluid.[34]

The cell count runs up to many thousands.

The blood count is not typical of influenza, there being a moderate leukocytosis usually. The blood culture is usually positive.

Duration is variable, somewhat slower than other forms, usually under two weeks, seldom over 20 days, but may exceptionally be extended.[35]

Prognosis.—Recovery is rare especially in infants. A small number, possibly 10 per cent., of older children may live, but they will be sadly crippled because of residual palsies.

Pneumococcus Meningitis.—This is the most frequent purulent type under consideration. The duration is shorter than usually found in the other types, frequently lasting only from three to six days. Infants in the early months are susceptible and most cases are under six months of age. The disease may precede, accompany or follow pneumonia, or be secondary to otitis media. Pneumococci are found in the blood cultures; older children may give evidence of a prodromal stage in which there will be headache, backache, dizziness, loss of appetite, vomiting; or these symptoms may develop suddenly with high fever, vomiting, restlessness, convulsions, drowsiness, delirium and unconsciousness. Petechiæ and herpes may occur.

At the brain base the pathologic changes are more pronounced in the posterior basilar regions. If pathologic changes are confined to the brain convexity, the usual symptoms of meningitis from basilar involvement will be absent, such as rigidity, respiratory and pulse changes, paralysis of the cranial nerves, and bulging fontanel. Pus and fibrin are unusually abundant in the exudate which may spread over the entire brain surface, the meninges covering the cerebellum are usually involved. In the few days which is the usual duration of the disease there may be insufficient time for hydrocephalus or encephalitis of severe nature to develop or for involvement of the spinal cord.

Spinal Fluid.—The spinal fluid contains large numbers of pneumococci. Differentiation from streptococci is not always easy. The fluid may be clear, serous, viscid, but it is usually gray, increased in amount, and forms a thick fibrinous sediment on standing. It becomes thickened and less abundant as the disease progresses. The cell count is usually very high and of the polymorphonuclear variety. Globulin and protein are greatly increased. Glucose is always absent. It is only by the bacteriologic examination of the spinal fluid and blood culture that the type of infection is determined. All types of pneumococcus have been found in the fluid, Types II and IV being the most common.[36] In all cases having pneumonia with secondary meningitis observed by Mitchell, the organisms were of Type IV.[37] The route of transmission is hematogenous.

Prognosis.—Mild cases have been known to recover, but the outcome in diffuse cases in children is always fatal. When improvement or convalescence occurs, the spinal fluid readily reduces Fehling's solution, which will aid one in predicting a favorable outcome.

Streptococcus and Staphylococcus Meningitis.—Infection with these purely pyogenic Gram-positive organisms is not common in infancy except in rare instances by extension from umbilical and spina bifida infection. It occurs more often in later childhood from foci of infection around the scalp, head injuries, erysipelas or otitis media, with mastoid abscess and sinus thrombosis. Meningitis may occur from infection of the middle ear during scarlet fever or from post-scarlatinal infection.

Erysipelatous meningitis is rare, but cerebral symptoms during the course of erysipelas of the scalp resembles meningitis.

The clinical and pathologic-anatomic picture is not very different from the other purulent forms. The onset is not sudden, but the temperature remains high, convulsions, stiff neck, positive Kernig and Brudzinski nearly always present. Hydrocephalus usually develops. The course is acute and fatal.

The spinal fluid is markedly cloudy and the organisms easily found. The usual evidences of inflammatory reaction and infection are found, such as increased pressure, high cell count of the polymorphonuclear variety and increased globulin.

In a death from streptococcus meningitis in which the original focus had been a purulent dacryocystitis the child developed a thrombosis of the cavernous sinus on the opposite side of the head.[38]

Colon Bacillus Meningitis.—One must carefully exclude toxic meningismus accompanying severe cases of pyelitis, in which organisms are not found in the spinal fluid.

Colon bacillus meningitis may occur during malnutrition, intestinal disturbances, colon bacillus pyuria, umbilical and spina bifida infections.

The writer has followed two cases in which the course was stormy, characterized by high temperature, convulsions, unconsciousness, opisthotonus, colon bacilli in spinal fluid; in one case the child is still living but with internal hydrocephalus, mental and physical retardation, strabismus; in the other case hydrocephalus of the communicating type increased until only a thin shell of cerebral cortex remained and death followed in three months due to the progressive and extreme emaciation.

The spinal fluid is brownish, thick, purulent, offensive and contains motile Gram-negative bacilli, both intra- and extracellular. The cell type is mostly polymorphonuclear.

The pathologic condition is illustrated in the findings at autopsy in the following case:

The brain of an 11-day old infant dying from colon meningitis showed a marked dilatation of the vessels of the pia mater, the arachnoid space contained an exudate made up of fibrin, leukocytes (mostly polymorphonuclear) and varying amounts of red blood cells.[39]

The symptoms and prognosis are not different from those of other types.

Typhoid Meningitis.—True typhoid meningitis must be differentiated from the serous or toxic. Meningitic symptoms are masked if typhoid fever co-exists especially if delirium is present. They may occur, however, after the febrile course has reached normal. Many bacilli, mostly intracellular, are found in the spinal fluid, and the fluid is slightly turbid, cells increased up to 1000, mostly polymorphonuclear, some phagocytes. The symptoms are acute and similar to those of other purulent types, but paralysis, convulsions and Kernig's sign are rare.

The brain at autopsy is covered both at the base and convexity with a diffuse, faint greenish-yellow or grayish purulent exudate, the dura may be involved, the ventricles dilated and full of turbid fluid, the meninges are cloudy, thickened, the brain convolutions pale and edematous.

There have been collected in recent years the histories of four cases of purulent meningitis in children where typhoid bacilli have been isolated from the spinal fluid. An interesting case was reported (Cole)[40] in a five-year old child who had a co-existence of typhoid and tuberculous meningitis.

Measles.—Meningitis of tuberculous origin or from suppurative otitis media may follow measles.

Mumps.—Leukocytosis may occur in the spinal fluid of mumps with or without meningeal symptoms. It is of a lymphocytic type and there may be at autopsy evidences of meningitis, but in the experience of clinical observers, it is rare.

Various Other Organisms.—A few cases have been reported caused by:

Bacillus lactic acid.
Bacillus subtilis.
Enterococcus meningitis.
Hemophilic streptothrix.
Micrococcus catarrhalis.
Micrococcus Florens.

Resumé of Spinal Fluid Findings

Although the gross appearance of the spinal fluid is similar for the purulent types, there will often be the following variations:

Pneumococcus—Pearl-gray fluid, sediment sinks to bottom of tube.

Streptococcus—Turbid, many cells and organisms, little fibrin.

Colon Bacillus—Dark-brown, offensive odor.

Influenzal Bacillus—Ground-glass, later greenish-yellow appearance, not many organisms.

Meningococcus—Yellowish or greenish sediment in a balloon-like mass.[41]

Lactic Acid Bacillus—Milk-white color, later curdled.[42]

Typhoid—Turbid, faint yellow tinge.

Though there is more or less difference in the gross appearance of the spinal fluid for the different types of purulent meningitis, the examination is not specific except for the presence of the causative organism in stained smears and cultures. In certain instances the fluid may rarely be clear and without clot formation. It is usually turbid and forms a clot on standing. It may be opaque, gray, yellow, green or brown. The polymorphonuclears are always in excess, the total cells may reach 2000 per cubic millimeter.

The butyric acid reaction (Noguchi) is found positive in all forms.

Sugar is reduced or absent.

The pressure is increased.

Prognosis.—Reported recoveries from suppurative meningitis should only be considered convincing if causative organisms have been found in the spinal fluid. Diffuse suppurative meningitis with demonstrable organisms is almost without exception fatal. Favorable operative treatment has been reported usually in those cases having a leukocytic fluid without organisms, or in cases where spinal fluid was not reported as examined or suggestive.

Treatment of the Purulent Meningitides.—Meningitis from hematogenous infection is not preventable, nor is any treatment as yet of avail.

Meningitis by otogenous extension should be prevented by removal of the primary focus through radical operation. In the developed case there should be reduction of the intracranial pressure. Prophylaxis should be inaugurated as early as possible.

Infection of the nasopharynx can frequently be avoided by removal of the adenoids. The tonsils should be removed if they are diseased or causing obstruction.

The suppurative meningitis which follows a brain abscess is a protective measure of nature in attempting to wall off the subarachnoid system; when the infection of the meninges becomes diffuse there is little hope unless the abscess is early drained by operation.[43]

The literature of suppurative meningitis contains references to operative procedures, craniotomy, laminectomy and drainage, spinal and cerebral puncture, irrigation of the cerebrospinal fluid circulating system, injection of specific and non-specific sera, vaccines. None of these measures have as yet been of proved benefit, because of the hopelessness of the cases especially since attempts have been made usually after the disease is well-advanced.

Irrigation of the subarachnoid spaces with Ringer's solution has been tried in three cases by Eagleton.[44] It is a highly technical and skilled method of surgical procedure, by which the solution is introduced through the spinal subarachnoid space and the outflow through a canula inserted into the cerebro-medullary cisterna. This lavage drains the subarachnoid system of pus. It is possible that this method may be developed by its early use into practical benefit in certain cases of otogenous infection.

In the influenzal type nothing specific has as yet been found curative. Repeated withdrawal of spinal fluid is usually attempted with the hope of effectually reducing the number of bacteria, the pus cells and the pressure. Homologous serum[45] or vaccins intraspinally or intravenously are worthy of trial but have thus far been usually of no proved effect. The Wollstein anti-influenzal serum has not yet been sufficiently tried to give evidence of benefit. The administration of hexamethylenamine and the establishment of permanent spinal drainage have not been successful.[46]

In pneumococcus meningitis daily lumbar puncture until the fluid becomes scanty has been tried.[47] This will sometimes relieve the nervous symptoms but is otherwise of no value. Anti-meningococcus serum is usually injected, because of the doubt regarding the diagnosis of the spinal fluid, and in the absence of any specific serum this is justified.

In streptococcus meningitis a polyvalent streptococcus serum may be used, but without expecting specific or even beneficial results.

An eight-year old child was reported cured following laminectomy and spinal drainage. The spinal fluid contained streptococci and leukocytes, and there had been a definite meningitis following mastoiditis. This case seems well proved so far as the diagnosis and recovery are concerned though the method of treatment is not in a single case to be accepted as the reason for the cure.[48]

Meningitis due to infection with staphylococcus aureus or albus is likewise fatal, but in the cases with less rapid course an autogenous vaccin might have some reason to prove valuable in this type.

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CHAPTER CLXXXIII

INTERNAL HEMORRHAGIC PACHYMENINGITIS OF INFANCY

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History.—The dura mater is involved in only one primary important pathologic process in infancy and that is the so-called internal hemorrhagic pachymeningitis. The early description of the disease notably by Virchow in the last century dealt almost entirely with its manifestation in adults where it was considered as dependent upon chronic alcoholism. As recently as 1904 the occurrence in infancy was thought by Finkelstein[1] as rare, at which time he reported a case in a seven months old infant. In 1905, Goeppert[2] reported three cases in children. In 1911, Finkelstein described six cases and showed pathologic specimens from five of these. By 1913, Rosenberg[3] was able to report over 38 cases, and had come to the conclusion that the disease is not rare in infancy. Cases began to be published in this country, Meigs,[4] in 1913, one case; Gordon,[5] 1914, two cases in children, in the discussion of which Spiller thought the disease more common in the young child than in the older.

Pachymeningitis was found in 3.9 per cent. of 6000 autopsies in children by Kowitz,[8] reported in 1914. Riley,[7] in 1915, described a case which died following the course of decomposition and internal hydrocephalus.

Schwartz[8] discussed the possibility of various sources of infection as the cause of the disease and reported in 1916 a case in which organisms of the meningococcus type were found.

In 1916, Feer[9] had seen five cases in Zurich within three years.

Rosenberg[10] in 1920 from his experience of many years classified the disease pathologically into two types, the traumatic newborn type and the later idiopathic progressive form, and reported the sequelæ in 10 children whom he had followed.

Finkelstein[11] in his 1921 edition of "Lehrbuch der Säuglings-Krankheiten" devotes eight pages to the subject, giving case reports, the symptomatology of three types, and a full discussion of the subject.

In 1923, Burhans and Gerstenberger[12] described five cases in their practice, the largest individual report thus far in America.

From the increasing knowledge of the disease, it is evident that in the future its recognition will be easier and more general, and the present somewhat confused ideas of the etiology and of the pathologic processes will be replaced by more intimate understanding of its true nature.

Other names for this disease have been offered as being more applicable to the pathologic condition than *pachymeningitis,* among them hematoma of the dura, cystic disease of the dura.

Definition.—Internal hemorrhagic pachymeningitis of infancy is a disease characterized by enlargement of the cranium from a pathologic increase in its contents, due to the occurrence of repeated hemorrhages and the formation of a serosanguineous transudate in and beneath the dural tissue, and the formation also of a subdural new-membrane which usually encapsulates the pachymeningeal fluid. The hemorrhagic process and the resulting increase in intracranial pressure cause certain pathognomonic signs and other brain symptoms which form a distinctive clinical picture.

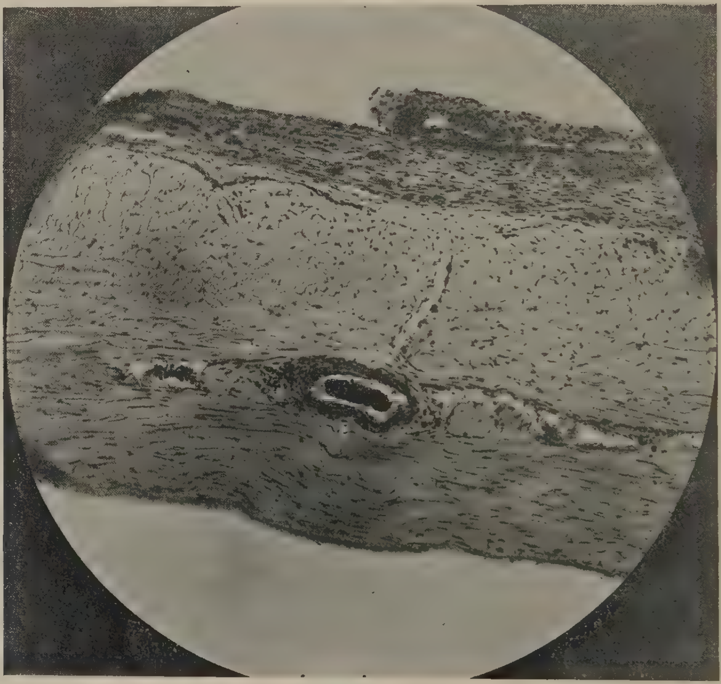

Fig. 65.—Pachymeningitis hemorrhagica interna. Photomicrograph of a section of the dura showing the new membrane formed upon the inner layer of the dura. (Photograph by Richard L. Sutton, Jr.)

Pathologic Anatomy.—It is well to recall that the *normal dura* covering the cerebrum is a thick fibrous membrane composed of two laminæ, an external (endocranial), and a smooth internal or meningeal layer (covered with endothelium). These laminæ are not separable except in certain areas where they form the venous sinuses. The dural tissue consists of white fibrous and elastic bundles, crossing each other in such a way as to form connective tissue spaces which are lymph channels and open into the subdural space. Numerous arteries and veins are found in the internal layer.

Location.—The convexity of the cerebrum in the regions of the anterior and middle fossæ is the usual site of the lesions, which form a cap-like cystic covering on one or both sides, more or less localized or extensive. In a case described by Holt[13] the autopsy showed a membrane covering not only the convexity but also the base. The posterior or cerebellar lobe of the cerebrum is seldom if ever involved. All authorities agree that the disease begins in the inner layer of the dura, and that it consists in a vascular proliferation of this portion of the dura with small and thin-walled capillaries from which oozing or larger hemorrhages take place. There results compression of the pia-arachnoid tissues and cerebral convolutions.

Proliferative and Transudative Processes.—A primary inflammatory stage was described by Virchow and others. This opinion is not held by the late writers who doubt that the process is inflammatory, because of the character of the fluid which is not that of an inflammatory exudate but is strictly a hemorrhagic transudate of proliferative origin.. Opinion is chiefly against the occurrence of a primary infective process in the dura although the hemolysis occurring in the hemorrhagic fluid has been recently ascribed to the presence of hemolytic streptococci (Rosenberg).

The nature of the process in infants varies with the etiology. If it has been caused by trauma, such as birth injury to the dura, the bleeding would either be persistent and soon cause death or if it subsides it would cause no further manifestation. So far as the complete picture of internal hemorrhagic pachymeningitis in older infants is concerned there is found spontaneous, recurrent and progressive hemorrhages and serous cystic formation. At autopsy the process shows a proliferation of the inner endothelial layer of the dura, going on to the formation of a new vascular membrane composed of many layers of delicate or firm connective tissue.

The ventricles are never involved and do not show any increase in their normal cerebrospinal fluid.

Repeated hemorrhages occur from the thin-walled newly proliferated capillaries and these fill and distend the connective tissue spaces. Organization of the clot takes place and layers are developed which go to form the new-membrane which acts to retain the fluid by a cyst-like wall. The older fluid is mostly a clear or yellowish serum, mixed with fresh blood from time to time. When the hemorrhage is single and circumscribed it is designated as "hygroma" of the dura mater. The essential local process does not differ in its pathologic anatomy from that described in adults.

Etiology.—Few plausible explanations of the origin of this disease have been given. None of the theories have as yet been proved. From the numerous diseases which have been described as forerunners of individual cases, it is probable that the causative agent is not always the same.

Hemorrhagic disease has been first thought of as the most logical cause. Rarely has any case been accompanied by hemorrhages elsewhere than within the cranium and in the eyes. Few observers have reported the coagulation or bleeding times and these were found normal (Burhans and Gerstenberger). The history of a hemorrhagic diathesis has been negative. Experimental production of intracranial hemorrhage and injection of blood into the dura of animals have not caused pachymeningitis.

Trauma as a cause does not explain the progressive and proliferative course, though the ultimate disposition of traumatic accumulation of blood inside the infant's cranium is not clearly understood. Pachymeningeal hemorrhage can clearly occur as an entirely different process from that found in pachymeningitis hemorrhagica interna.

Cases appear in infants who have been previously in a good state of nutrition while there are instances of the disease reported in undernourished infants. If athrepsia be a predisposing cause it must be rare, for the clinical picture of pachymeningitis is not found as a feature of low nutritional states. Scurvy and rickets have been offered as explanations of the disease, but without proof. In either of these cases there should be response to appropriate treatment.

When we examine the history of these infants as pertaining to infections, we find only a few who have lues or have had pertussis. Finkelstein and Rosenberg offer the best explanation. They found that outbreaks of hemorrhagic rhinitis in the infants at the Berlin Kinderasyl were followed in from two to four months by the occurrence of several cases of pachymeningitis hemorrhagica interna. The rhinitis may have been due to lues but in most casss to nasal diphtheria as proven by culture. Their theory, not yet proven, is that thrombotic intermediary changes take place in the ethmoidal and ophthalmic vessels and then in the cavernous sinuses, which cause a passive congestion in the dura and resultant hemorrhage.

Symptomatology.—Cranial enlargement of moderate size is characteristic of pachymeningitis hemorrhagica interna. This develops usually slowly from the increase in intracranial pressure due to the progressive formation of fluid and the pachymeningitic tissues. A rare and severe type, however, is described as an *acute meningitic form* in which there is not time for marked enlargement of the skull, though the sutures are somewhat separated and the fontanel tense and wider than formerly. There are the usual cerebral symptoms of meningitis, beginning with vomiting which is repeated. Convulsions, stiff neck, muscle hypertonus, increased knee-jerks, stupor; eye-symptoms may be present including strabismus, and eye-ground changes with retinal hemorrhage, congested veins and paleness of the papillæ. Coma develops and in a few days death occurs.

An *intermediate type* due to intra-cranial pressure is described by Finkelstein. This is more common than the acute meningitic type, but the symptoms are much the same and the outcome not so fatal. There are mild symptoms of pressure which may disappear, or the symptoms persist and the disease merges into the chronic form.

Pachymeningitis hemorrhagica interna is more often a *chronic* disease lasting several months, with insidious development. The most usual symptoms are:

Slow and moderate enlargement of head (external hydrocephalus).
Gradual separation of sutures.
Distention of the fontanel.
Restlessness.
Intelligence and nervous condition may or may not be affected.
Eye-grounds show punctate or linear hemorrhages in retina and around the papillæ and the veins tortuous.
Usually from one to three degrees of fever.
Reflexes increased.
Slight muscle hypertonus.
Decline in weight occurs.
Finally the child improves and may become normal, but usually is impaired.

These cases frequently succumb to pneumonia or other intercurrent disease.

Thus it will be seen that the general symptoms are not definite for this disease alone. There may be other symptoms from cortical irritation and basal irritation (Rosenberg).

Diagnosis.—Pachymeningitis comes into question whenever there is any pathologic increase in the head circumference especially of moderate amount. Other diseases which must be differentiated are luetic hydrocephalus, so-called rachitic enlargement of head, meningismus and tuberculous meningitis. The history of previous illness and infection, the existence of cerebral symptoms, spasticity, increased reflexes and failing nutrition are of value but the specific diagnosis rests upon the findings from fontanel puncture, eye-ground examination, and of less importance lumbar puncture.

Fontanel Puncture.—A short bevelled trochar is pushed through the scalp at a sufficient distance (at least 2 cm. to either side of the longitudinal sinus in the region of the anterior fontanel). Immediately upon piercing the dura, a hemorrhagic or serous fluid, clear, yellowish or brown, escapes under pressure. When this fluid is allowed to stand in the tube a sediment of red cells is formed. No clot nor fibrin pellicle forms. The supernatant fluid is clear or yellowish, or reddish-brown, the result of hemolysis. If recent hemorrhage has occurred, the cystic fluid will be colored thereby. The fluid contains little albumin usually, and is sterile. The procedure of puncture is a simple one.

Ophthalmoscopic examination should be performed in every suspected case, as hemorrhages in the fundus are valuable evidences, though not always present (in from 30 to 80 per cent. as reported by various observers). Retinal hemorrhages in one or both eyes are diagnostic as a part of the symptom complex. The disc may show a congestion or a neuritis. Occasionally there will be superficial hemorrhages in the eyes—conjunctivæ and lids.

Lumbar Puncture.—Pressure is increased in all reported cases. The withdrawal of the available spinal fluid does not reduce the tension of the fontanel nor is there usually evidence of blood in the cerebrospinal fluid unless the pachymeningitic fluid is incompletely encapsulated (twice in the 38 cases of Rosenberg).

Course.—The disease is usually encountered in the first year of life, the majority of cases occurring between the ages of three to five months. The disease lasts from one to six months. Recovery is slow accompanied by a gradual cessation in the intracranial pressure and retinal hemorrhages, a closure of the sutures, and ossification of the fontanel—which sometimes remains depressed.

Probably a long time is required for the complete disappearance of the organized tissues and new-membrane in the affected dural area.

Prognosis.—The acute and severe types are nearly always fatal. Those without severe meningeal symptoms and running a chronic course have a good chance for recovery so far as life is concerned, and in some cases without damage to the nervous and mental functions. It is common, however, to find a permanent impairment of the intelligence. Disturbances of vision and ocular palsies are not infrequent sequelæ.

Rosenberg made 10 late examinations to determine the outcome of the disease and found that there were two idiots, three imbeciles, one neuropathic child, one stutterer, one bedwetter, two normal. Optic atrophy led to severe visual disturbance in two children and one had facial palsy after eight years.

Persistent facial paralysis, blindness in one or both eyes, and hydrocephalus have been observed. Pneumonia is the most common cause of death. Secondary infection and abscess of the pachymeningeal tissues, the leptomeninges and the cerebrum have occurred.

Hematogenous infection may localize in the pachymeningeal areas as a result of lowered resistance.

Treatment.—Recovery is spontaneous, rather than the result of any treatment thus far tried. Feer believes that the results from fontanel puncture are good. Repeated aspiration may temporarily relieve the pressure symptoms, but it would seem to have no effect upon the proliferative and hemorrhagic process. Fontanel punctures should be made on both sides. Finkelstein has seen patients recover with the weekly injection of gelatin solutions, but he has had also unsuccessful cases, and he does not feel sure that there is any mechanical or specific value therefrom. Caution should be used in the aseptic technique of all punctures, as infection may be caused.

General measures will include the proper feeding and care of the infant in the improvement of nutrition; suitable blood transfusions may be tried, and anti-syphilitic drugs if indicated.

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CHAPTER CLXXXIV

ANATOMY AND PHYSIOLOGY OF THE SPINAL CORD AND VARIOUS AFFECTIONS OF THE SPINAL MENINGES AND CORD

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ANATOMY AND PHYSIOLOGY OF THE SPINAL CORD

From a neurologic point of view the differentiation of the spinal cord from the medulla, pons and basal ganglia is a more or less arbitrary procedure, only to be tolerated as a matter of descriptive convenience.

On this basis therefore the spinal cord may be considered as that part of the central nervous system that extends from the lower portion of the pyramidal decussation, in the medulla, to the filum terminale and the cauda equina, in the lowermost portion of the spinal canal. In its external skeletal relationship it is that part of the central nervous system that extends from the foramen magnum to the lower border of the first lumbar vertebra.

The termination of the cord proper is in the form of a cone-like point, the conus medullaris, from which there descends a filamental prolongation, the filum terminale, which, with the roots of the lumbosacral nerves, forms the cauda equina.

The filum terminale is a prolongation of the pial covering of the cord proper, and contains in its upper portion, just below the conus, more or less rudimentary gray and white substance continuous with that of the conus.

At about the level of the third sacral vertebral segment this filamental extension of the pia becomes fused with the dura-arachnoid to form a ligamental attachment that blends with the periosteum and is inserted into the coccyx.

The cord is not quite cylindrical in shape, being somewhat less in its anteroposterior than in its transverse diameter.

It is nearly divided into symmetrical halves by the anterior and posterior median fissures. The anterior fissure is broader but much shallower than the posterior. It is penetrated by the pia and contains the anterior spinal artery and its branches. The narrow posterior fissure penetrates the cord for more than half its diameter. It is filled with connective tissue to which the pia is attached but which it does not penetrate. It contains the ramifications of the posterior spinal arteries. In the anterior and in the postero-lateral regions of the cord throughout its extent are situated the roots of origin of the 31 pairs of spinal nerves, namely, eight cervical, 12 dorsal, five lumbar, five sacral, and one coccygeal. At the level of origin of the roots of the cervical nerves forming the cervical and brachial plexuses, and extending as far as the second dorsal vertebra, the cord is enlarged, chiefly in its transverse diameter (intumescentia cervicalis). It reaches its maximum opposite the fifth cervical intervertebral space. In the dorsolumbar region, corresponding to the origin of the roots of the lumbosacral plexus there is another enlargement (intumescentia lumbalis). It commences at about the tenth dorsal vertebra, reaches its maximum at the twelfth, and then tapers off into the conus.

These enlargements correspond to the greatly increased aggregations of nuclei, and the outgoing and incoming fibers, necessary for the innervation of the extremities.

The spinal nerves emerge from the cord in fan-like bundles, which gradually converge and form the single trunks of the individual roots. Each pair is composed of an anterior motor and a posterior sensory division, which unite to form the mixed sensori-motor trunks from which the peripheral nerves are formed. These roots are sheathed with all three layers of the spinal meninges. The lumbar and sacral nerves are massed together within the lower lumbar and sacral portion of the vertebral canal, forming the cauda equina. The entire cord is invested by a double layer of pia, the inner layer of which is continuous with the pia of the brain. It is intimately adherent to the cord. Laterally the pia, covered with arachnoid, is extended from each side of the cord in the form of a longitudinal band, and is attached to the dura by tooth-like prolongations, opposite each vertebra, from the atlas to the first lumbar vertebra inclusive. This is the ligamentum denticulatum. It divides the subarachnoid space into two parts, an anterior and a posterior, filled with cerebrospinal fluid. These spaces have free communication through the intervals between the tooth-like processes of the ligament.

The subarachnoid space of the cord communicates directly with the subarachnoid space of the brain through the foramen magnum.

The arachnoid is a delicate membrane continuous with that of the brain, attached internally to the pia along the posterior portion of the cord by fibrous bands, but elsewhere separated from it by the subarachnoid space filled with cerebrospinal fluid. Externally the arachnoid lies close to but nowhere in contact with the dura, a small amount of cerebrospinal fluid intervening.

The dura sac of the cord hangs suspended from the margin of the foramen magnum and the bodies of the three upper cervical vertebræ. While it is continuous with the dura of the intra-cranial space, it differs from it in several respects. Except for its attachments in its upper portion it hangs free from the walls of the vertebral canal, and nowhere possesses any periosteal function, as it does for the inner plates of the cranial bones. It is separated from the vertebral wall by areolar and fatty tissue and the vertebral venous plexus. Both surfaces of the dura are covered by endothelium. Within the cord is a central canal, commencing in the fourth ventricle in the medulla, and extending throughout the length of the cord to the conus, where it becomes somewhat dilated, forming the so-called terminal ventricle, then narrows gradually and ends blindly about half way through the filum terminale. It is lined with ciliated columnar cells. It varies somewhat in form and size in different parts of the cord. It should be just visible to the naked eye.

It is sometimes more or less obliterated, especially in the cervical cord.

During early fetal life the spinal cord completely fills the vertebral canal and its segments correspond to the respective vertebræ opposite to which they are located. Owing to the rapid lengthening of the vertebral column, even during intra-uterine life, it reaches at birth only to the bottom of the third lumbar vertebra, and the cord segments no longer correspond to the vertebra. In later childhood, by the twelfth year, and in adult life it terminates opposite the lower part of the first lumbar vertebra and the discrepancy between the spinal segments and the vertebræ is still further accentuated. On account of this difference in the relation of the cord segments to the vertebral canal in infancy and in later childhood, the spinal roots make their exit from the vertebral canal relatively sooner in the early period of life than in later childhood. This is especially true of the lower dorsal and lumbosacral roots. Their course is less oblique and their point of exit nearer the level of the cord where they originate than is the case in later years.

As will be seen later this fact is of practical importance in segment localization and its relation to the external topography of the vertebral column in connection with surgical interference in diseases of the spinal cord.

Another feature that is conspicuous, on macroscopic inspection of the infantile cord, is the relatively slight differentiation of the cervical and lumbar enlargements, in size and form, from the rest of the cord.

Histologically and physiologically the essential characteristic of the spinal cord, as of the rest of the nervous system at birth, is its immaturity. The spinal cord at birth is, however, relatively better developed than any other part of the nervous system. Even in fetal life it is called upon to answer certain functional demands of automatic character at a period when the rest of the nervous system is for the most part functionally unawakened. Consequently at birth it is in a state of development that renders it capable of responding more or less perfectly to the primitive demands of the first few days of life.

So rapidly however do these demands multiply in the early weeks of life, and so active is the resulting histologic growth, that the cord has doubled its weight by the end of the fifth month and trebled it by the end of the first year. With the termination of infancy, at the end of the second year, it has quadrupled its weight at birth.

The arterial supply of the spinal cord is derived mainly from the vertebral arteries. From the viewpoint of a cross segment of the cord, the distribution may be roughly divided into three fields for each lateral

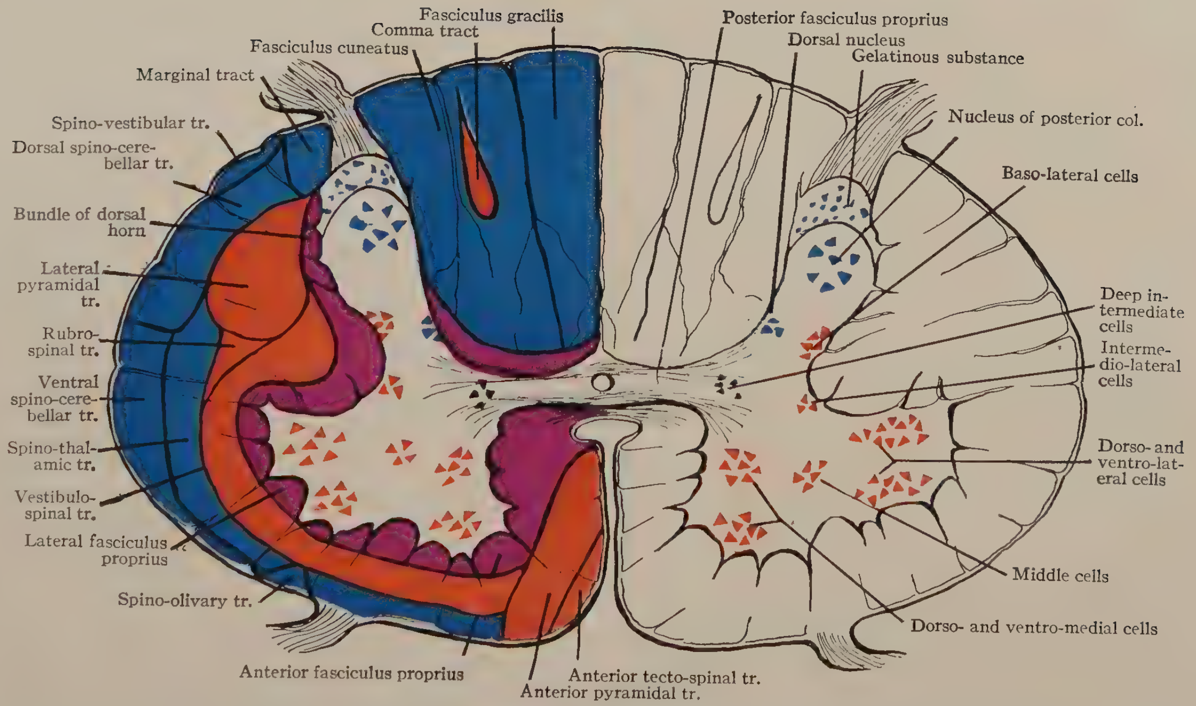


Fig. 66.—Diagrammatic representation of a cross-section of the cord at the level of the sixth cervical segment. (From Santee, Anatomy of the Brain and Spinal Cord, Blakiston, 1915.)

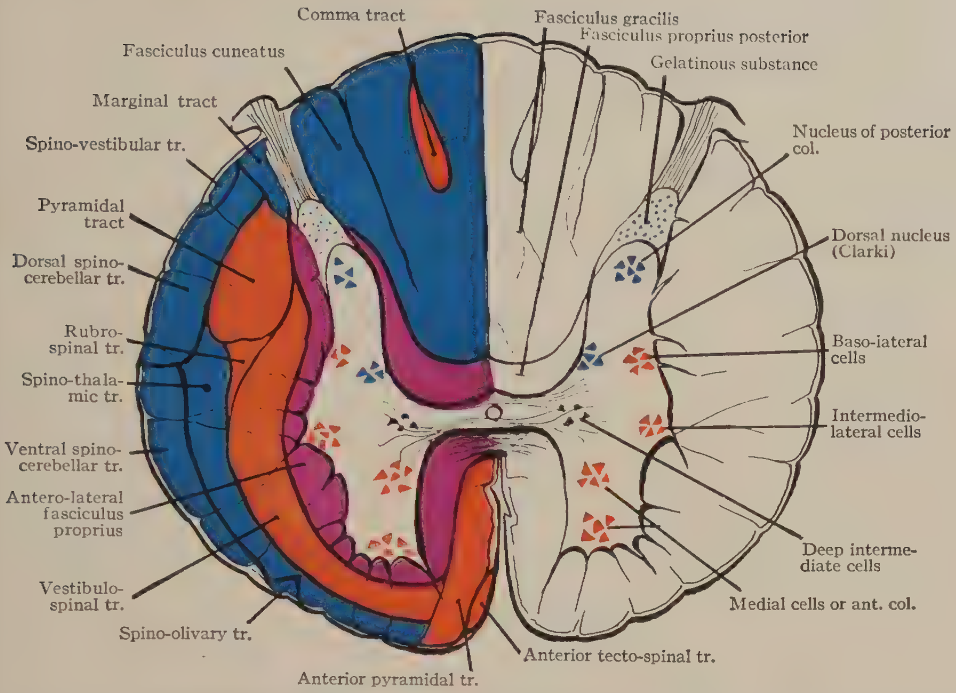
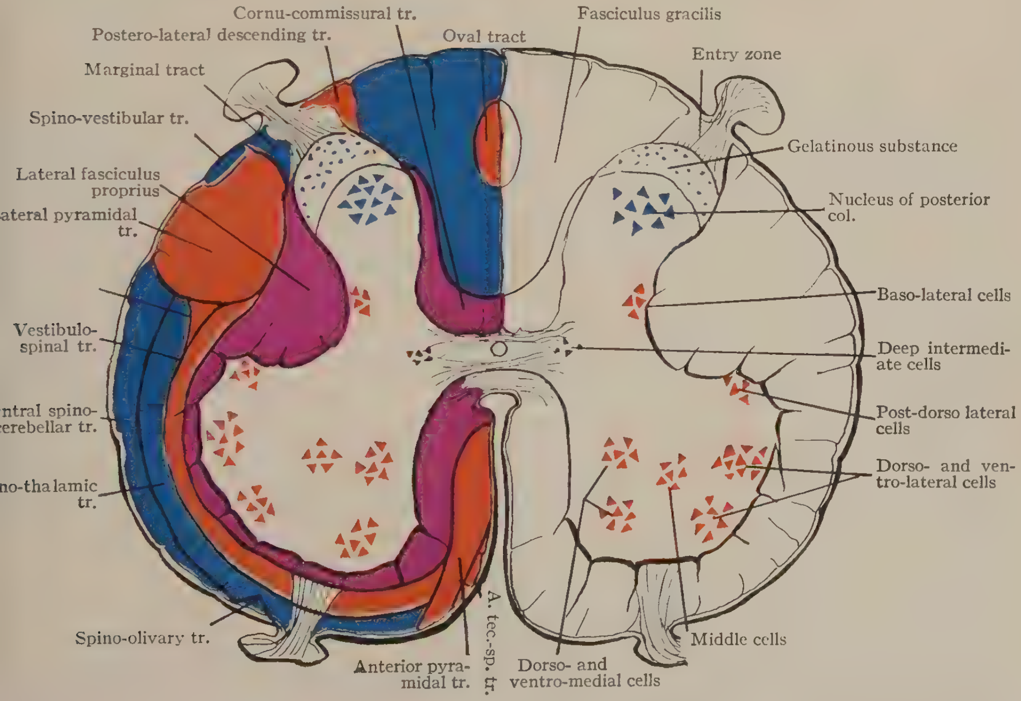


Fig. 67.—Diagrammatic representation of a cross-section of the cord at the level of the sixth thoracic segment. Red tracts are descending; blue tracts are ascending, and purple tracts are composed of both ascending and descending fibers. (From Santee, Anatomy of the Brain and Spinal Cord, Blakiston, 1915.)



g. 68.—Diagrammatic representation of a cross-section of the cord at the level of the third lumbar segment. (From Santee, Anatomy of the Brain and Spinal Cord, Blakiston, 1915.)

half of the cord. The anterior field, comprising most of the area of the anterior columns, and all of the gray matter of the anterior and central portion of the cord, is supplied from the anterior spinal artery, which, originating from the vertebral arteries within the cranial cavity by two trunks which join about the level of the foramen magnum, continues down the entire length of the cord at the entrance of the anterior median fissure and beneath the pia. The posterior field is supplied by the posterior spinal arteries, also originating in the vertebral arteries. These arteries, one on each side, run down the length of the cord close to the posterior spinal roots. They supply the posterior columns, the posterior root-zones and the posterior gray substance of the cord. The lateral field lies between the anterior and posterior spinal roots, and is supplied by a plexus made up of the anastomosing branches from the anterior and posterior spinal arteries, and from the vertebrals in the upper, the intercostals in the dorsal, and the lumbar, ilio-lumbar and lateral sacral arteries in the lower region of the cord. The anterior and posterior spinal roots are supplied by branches from the posterior spinal arteries in the various regions of the cord. From the root branches are obtained the anastomosing branches mentioned. It will be noted from the above described arterial supply that there are regions of the cord that are supplied solely from the anterior spinal artery, others supplied only by the posterior spinal arteries, and others deriving their supply from both sources, as well as from the extraspinal sources mentioned.

While the arterial branches anastomose intricately before entering the cord, they do not do so after their entrance. They are therefore terminal arteries.

It must not be inferred that the arterial fields outlined are sharply differentiated, for there is of course some overlapping, but in a general way such a conception of the arterial supply holds good, and bears an interesting relationship to the pathology of many spinal diseases.

Numerous small veins emerge from the surface of the periphery of the cord and along the spinal roots and, uniting, overspread the entire surface of the cord in a dense plexus lying between the pia and the arachnoid. At the upper part of the cord they unite in two or three small veins and empty into either the inferior cerebellar veins or the vertebral veins. The plexus communicates freely with the extradural internal vertebral plexus and via this route drains into the vertebral, intercostal, lumbar and sacral veins. The spinal veins are devoid of valves.

The cord is composed of gray and white matter. The gray matter occupies the central portion of the cord. It is made up of innumerable aggregations of ganglia cells embedded in a mass of neuroglia, supported by connective tissue derived from the pial covering of the cord, of axones and dendrites of these cells, and of white fibers traversing the neuroglia on their way to these ganglia cells or to more distant parts of the nervous system. These cells of the gray matter of the cord constitute subordinate power and relay stations for the number-less nervous impulses necessary for the maintainence of the vegetative, automatic and voluntary functions of life.

Within this gray matter originate the motor impulses that produce all of the spinal reflexes and at certain levels are situated special centers presiding over special functions, such as pupillary dilatation, diaphragmatic action, bladder, rectal and genital functions.

To facilitate localization the gray matter of each lateral half of the cord may be considered as consisting of three parts the anterior and posterior horns and the central portion. In the anterior horns are located aggregations of ganglia cells in which is generated the motive power of the body. This motive force includes not alone that which is under voluntary control, but also sympathetic motor activity, as well as manifold reflex acts, either partially or not at all under the control of the will, and governing automatic functional activity of many kinds. In the posterior portion of this gray matter of the cord are located collections of ganglia cells that constitute relay stations for the reception and further transmission of centripetal impulses from the periphery.

The central gray portion is occupied by association and commissural neurones and reflex relay neurones, and through this portion pass the sensory fibers from the posterior roots on their way to the antero-lateral tracts. In the outer part of this portion of the gray matter is a collection of cells, the intermedio-lateral column. This column of cells supplies involuntary muscles and glandular function. It is a visceral nucleus controlling vasomotor, trophic and secretory activity, and in it are located, at different levels of the cord, special centers governing organic involuntary motor function.

A glance at the diagrams (Figs. 66–68) will show the approximate location of these centers as seen in cross-section of the cord.

Outside of the gray matter is situated the white substance of the cord, a dense mass of cables connecting the cord with the brain and with the periphery of the body, and the different levels of the cord with each other. The association and commissural tracts are located close to the gray matter. They bring the different cord levels into interrelation with one another, the association tracts joining different levels of the same side, the commissural tracts joining different levels of opposite sides of the cord.

The white substance of the cord compared with the gray matter is relatively better developed at birth, but only relatively so.

The requirements of extra-uterine life demand constant increase of these white fibers, both in the long spino-medullary and cerebrospinal tracts and in the shorter association and commissural tracts. Therefore, active formation of axis cylinders and their subsequent myelinization is going on throughout the period of infancy and childhood in the cord as elsewhere in the nervous system.

Many of the white fibers of the cord, existing at birth, are not myelinized and this process takes place in the early months of extra-uterine existence.

Thus, contrary to the views that at one time prevailed, it has been definitely shown that the cerebral portion of the pyramidal tracts is developed as far as the cervical enlargement of the spinal cord as early as the seventh month of the fetal life,* and that this portion is completely myelinized shortly before birth. During the last two months of fetal life, while the sheathing of these tracts in myelin is taking place, the rest of the pyramidal tract is being extended down the cord to the lumbar region, but the myelinization of these fibers occurs during the first year of *extra-uterine* life.

The white substance of the cord is divided, for convenience of descriptive localization, into an anterior, a lateral and a posterior portion, and the columns that pass through these regions are designated accordingly, the anterior, antero-lateral, lateral and posterior columns (funiculi).

These columns are made up of various tracts (fasciculi) as indicated in the diagrams. These tracts are composed of ascending or of descending, or of mixed, ascending and descending fibers.

A brief description of some of these tracts is unavoidable in order to make clear the pathologic basis for the clinical phenomena of spinal disease. On the other hand, it is desirable to avoid, as far as possible, the inclusion in a clinical treatise of confusing details of the course of tracts of which the exact function is not yet fully demonstrated, however interesting this may be from an anatomic and physiologic point of view.

In the following descriptions each fasciculus of one-half of the cord is described, it being understood of course that its counterpart exists in the opposite half of the cord and follows a similar course. The description of the location of these fasciculi is from the viewpoint of a cross-section of the cord. The diagrams (Figs. 66–68) should be consulted in connection with the text.

Funiculi Anteriores.—Fasciculus Tectospinalis Anterior.—On each side of the anterior median fissure of the cord there is a small bundle of fibers that originates in the superior quadrigeminal body of the same side and passes downward beneath the aqueduct of Sylvius, through the pons and the medulla to the eighth cervical segment of the cord where for the most part they terminate in the ciliospinal nucleus. This tract is part of the reflex arc for pupillary dilatation. A lesion of this tract breaks the arc and destroys the reflex dilatation of the pupil to diminishing light. Some fibers from this fasciculus may be traced well down to the lower cord.

Fasciculus Pyramidalis Anterior.—Immediately adjacent to the tract just described is a bundle of fibers originating in the cerebral motor cortex and descending uncrossed through the internal capsule, crus, pons and medulla to this portion of the cord where some, if not all, cross in the anterior commissure and terminate about the cells of the anterior gray matter of the cord from the cervical to the lumbar enlargement. Lesions of this tract will be considered in connection with lesions of the larger lateral or crossed pyramidal fasciculus of the lateral region of the cord.* They constitute from 10 to 20 per cent. of the entire pyramidal fasciculus, according to Marchi.

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* Van Gehuchten, Nevraxe, iii, S. 263 (1901); Rev. de Neurol., S. 65 (1897); Jour. de Neurol. (1896).

Van Gehuchten, Faisceau pyramidal et maladie de Little. Jour. de neurologie (1896); Maladie de Little et rigidite spasmodique spinale des enfants nes avant terme. Rev. de neurologie (1897), p. 65.

Brissaud, Lecons sur les maladies nerveuses. Paris (1895), p. 108, etc.

Funiculi Anterolaterales.—Fasciculus Anterolateralis Proprius. Adjacent to the fasciculus just described, and between it and the gray matter of the anterior horn, and in immediate proximity to it, is the anterior portion of an extensive fasciculus of mixed ascending and descending fibers that extends entirely around the gray matter of the anterior horn and lateral regions of the cord nearly to the posterior nerve roots. This fasciculus is made up of the ascending and descending axones of cells of the adjacent gray matter, and forms for the most part short association and commissural tracts between different levels of the gray matter of both sides of the cord.

Fasciculus Vestibulo-spinalis.—External to the fasciculus just described, and extending from the anterior to the lateral pyramidal fasciculus, is a tract originating in the vestibular nucleus of the pons and medulla (Deiter's nucleus) and terminating in the gray matter of the anterior portion of the cord. Through the vestibular nucleus it connects with the cerebellum.

It is an important part of the complex mechanism presiding over equilibrium and coördination, especially in relation to locomotion. It is a descending tract.

Fasciculus Spino-thalamicus.—External to the vestibulo-spinal fasciculus, and extending like it around the cord from the anterior to the lateral pyramidal tract, is the spino-thalamic tract, composed of bundles of ascending sensory fibers conveying sensations of pain, temperature and tactile localization and a certain number of fibers transmitting sensation for light touch and pressure touch (contact perception). This fasciculus is made up of fibers that are the axones of cells in the posterior and central gray matter of the other side of the cord, and have crossed in the anterior gray and white commissures to this side.

Impulses reach these cells by fibers coming in through the posterior roots and root-zones and terminating about these nuclei of the posterior gray matter. These nuclei therefore are the nuclei of origin of this fasciculus. It degenerates upward only when the lesion is in or proximal to these nuclei. In other words, this tract does not degenerate in destructive lesions of the ganglia of the posterior roots or in lesions of the posterior roots themselves.

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* Sections of the cord above the lesion in certain instances of transverse lesion of the cord seem to indicate that there are some ascending fibers in this region. Whether the ascending degeneration in such lesions is merely the implication of short association fibers or involvement of more extended fasciculi is not clear.

The tract ends in the lateral nucleus of the optic thalamus. These last three fasciculi are all pierced in their anterior portion by the anterior nerve roots, dividing them into an anterior and a lateral portion.

The anterior part of the spino-thalamic fasciculus is supposed to convey only sensation for light touch and pressure touch.

Small circumscribed lesions of this tract may produce dissociated sensory disturbance, a loss of perception of one form of sensation with other forms intact, although this is more commonly due to a lesion of the central gray matter.

A lesion involving the whole of this tract abolishes sensation of pain and temperature in the other half of the body below the level of the lesion. Tactile sensation and muscle sense are transmitted through the posterior columns and would not be abolished by any such lesion, although some disturbance of tactile sensation might be evident through involvement of the tactile sense fibers that are thought to pass through the anterior portion of this fasciculus.

Funiculi Laterales.—Fasciculus Spino-cerebellaris Ventralis.—Occupying the periphery of the lateral region of the cord, and external to the tract just described, is the ventral cerebellar fasciculus, extending throughout the entire length of the cord. It accompanies the spino-thalamic fasciculus as far as the pontine nucleus of the trigeminal nerve. These two tracts are commonly known as Gower's tracts.

The ventral cerebellar fasciculi from each half of the cord decussate in the cerebellum and terminate in the superior vermiform process. These fasciculi are supposed to carry ascending sensory impulses to the cerebellum and to serve as the centripetal portion of a reflex mechanism of coördination. Whether or not conscious sensory impulses are ever received in the cerebral cortex, via this route, is not definitely known, although it is theoretically possible.

Fasciculus Spino-cerebellaris Dorsalis.—Occupying the rest of the lateral periphery of the cord, in the cervical and dorsal region, but not in the lower lumbar segments, is another ascending cerebellar tract, ordinarily known as the direct cerebellar tract (Flechsig). It is situated immediately posterior to the ventral cerebellar tract just described, and extends back nearly to the posterior spinal roots, being separated from the latter by the marginal tract (Lissauer).

The fibers of this dorsal cerebellar fasciculus arise in the cells of the dorsal nuclei at the base of the posterior horn of the spinal gray matter ("Clark's column") and run directly to and terminate in the cortex of the superior vermiform process of the cerebellum.

It is now thought that this fasciculus transmits sympathetic impulses, especially from the viscera. It is not probable that this is so.

Fasciculus Spino-vestibularis (Horsley-Thiele).—External to the extreme posterior portion of this fasciculus is a small tract that consists of ascending fibers arising in the lumbo-sacral cord and ending in the vestibular nucleus. They probably are connected with the function of equilibration.

Fasciculus Pyramidalis Lateralis.—Immediately beneath the direct cerebellar tract lies the lateral pyramidal tract, composed of the crossed pyramidal fibers descending from the cerebral cortex to the cells of the anterior gray matter of the cord. In the lumbar cord this tract extends to the periphery of the cord on account of the disappearance of the direct cerebellar tract in that region. The pyramidal system transmits voluntary impulses from the cerebral cortex to the ganglia cells of the lower motor segment, and initiates in these cells impulses that result in muscular activity, as well as exercising a certain control (inhibition) over any tendency in the lower motor segment to irresponsible action.

Involvement of the pyramidal system by trauma or disease furnishes some of the most common and important clinical pictures seen in the entire domain of the nervous diseases of childhood, as well as of adult life. Most of these in childhood have to do with lesions of the cerebral portion of the pyramidal system.

Among the affections in which the spinal portion of the pyramidal system is or may be involved, either alone or in conjunction with the cerebral portion, are the following: Lateral and amyotrophic lateral sclerosis, hereditary ataxia, hereditary spastic paraplegia, residual of acute myelitis, meningomyelitis (syphilitic or other forms), multiple sclerosis, tuberculous spondylitis, and occasionally tumors or abscess in or adjacent to the cord.

Dislocation of vertebræ, provided the cord is not crushed, may also be accompanied by a spastic paraplegia, due to pressure on and consequent interference with conduction through these tracts, as in the child whose case is referred to in the section on localization.

Fracture of a transverse process of any one of the lower cervical or the dorsal vertebræ, if it is sufficiently displaced to press upon the lateral region of the cord, may give rise to clinical signs of unilateral pyramidal lesion—spastic paralysis of one leg. The characteristic clinical features of involvement of this tract are a loss or diminution of the power of volitional control of muscles, muscular spasm, the appearance of certain abnormal reflexes never seen except in lesions of this tract. The normal superficial or cutaneous reflexes are abolished in these lesions.

All of these phenomena occur below the level of and on the same side as the lesion, when the latter is situated in the spinal cord.*

Fasciculus Rubro-spinalis.—Immediately anterior to the lateral pyramidal tract, and deep within the lateral white substance of the cord, is a tract of descending fibers emanating partly from the red nucleus beneath the optic thalamus and partly from the thalamus itself and terminating in the anterior gray matter of the cord. The fasciculi from the red nucleus cross in the tegmental decussation. They are believed by Horsley to transmit impulses having to do with coördination, especially of the act of locomotion. It is sometimes referred to as Monakow's tract.

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* A phenomenon that I have several times seen, although it is rare, is an exaggeration of the deep tendon reflexes and the appearance of abnormal reflexes (Babinski, Oppenheim, etc.) on the side opposite the site of the lesion. See Fairbanks, The contralateral plantar reflex, Rev. Neur. and Psych. (1911), p. 225.

Santee states that the thalamic portion of this fasciculus is probably the carrier of reflex impulses.

Fasciculus Marginalis (Lissauer's Tract).—This tract of ascending fibers lies just anterior to the entering bundles of the posterior nerve roots. It is formed from fibers from the lateral bundle of these roots. Its function is as yet unknown. Fasciculus spino-olivaris: Just posterior to the anterior nerve roots is another small ascending tract of which the function is also unknown.

Funiculi Posteriores (Columns of Goll and Burdach).—Placed between the posterior root-zones and the posterior median fissure of the cord are two columns consisting for the most part of ascending fibers, the fasciculus cuneatus (Burdach) and the fasciculus gracilis (Goll).

Fasciculus Cuneatus.—This tract first appears in the cord in the mid-dorsal region and, continually enlarging by the accession of fibers from the posterior roots, passes upward between the posterior root-zone and the fasciculus gracilis and terminates in the cuneate nucleus in the medulla. It is an uncrossed tract.

Fasciculus Gracilis.—Appearing in the lowermost segment of the cord, this tract runs up between the column of Burdach and the posterior median fissure, and terminates in the nucleus gracilis on the same side of the medulla. It is, therefore, also an uncrossed tract.

Below the tenth dorsal segment it forms nearly the whole of the posterior column.

The nuclei of origin of both of these fasciculi lie in the spinal ganglia on the posterior spinal roots and are, therefore, outside of the cord. These fasciculi are the axonic processes of these ganglionic cells whose dendrites are the peripheral sensory nerves.

In addition to these ascending fasciculi of the posterior columns there is within the cuneate fasciculus, as far down as the ninth dorsal segment, a tract of descending fibers often referred to as Shultz's comma tract. It is formed by the descending branches of the Y-shaped divisions of the posterior root fibers. Below the ninth dorsal segment it shifts its position and lies next to the septum. The fibers of this tract descend through two or three segments of the cord. Their function is unknown. Immediately adjacent to the gray matter of the posterior commissure is a mass of both ascending and descending fibers, the fasciculus proprius posterior, an association and commissural tract connecting different levels of the cord, similar to the tract in the antero-lateral field.

The posterior columns transmit impulses of tactile and muscle sense.

Lesions limited to these columns produce disturbances of the sense of touch and of the function of coördination. The clinical picture therefore is that of diminution of tactile perception—hypesthesia—or of loss of the same—anesthesia, loss of perception of form, size and weight of objects, and loss of perception of vibration, loss of the sense of position of the limbs, ataxia and disturbances of equilibrium.

They are rarely involved without coincident involvement of the posterior roots and root-zones. Consequently to the above symptoms there are usually added subjective and objective disturbances of pain and thermal sensations, loss of reflexes, both superficial and deep, and often visceral crises.

Among the affections in which these columns are implicated are tabes, hereditary ataxia, postero-lateral sclerosis, meningo-myelitis, and tumors in or adjacent to the cord. In all of these affections the posterior roots and root-zones are usually involved, and not infrequently the spinal ganglia as well.

It is important to remember that all of the fasciculi of the cord degenerate from any given lesion in a direction away from their nuclei of origin, along the axis cylinders—axones of their nuclear cells. Thus for instance when the pyramidal fibers in the internal capsule are destroyed by vascular or other lesions, the entire pyramidal fasciculus below the lesion degenerates downward throughout the entire extent of the cord, for the nuclei of origin of this fasciculus lie in the cerebral cortex, above the lesion. When, however, a lesion occurs in the posterior columns these degenerate from the lesion upward to the medullary nuclei where these fasciculi end, for their nuclei of origin lie below the lesion, in the ganglia of the posterior spinal roots. When the cells of Clark's column are destroyed there occurs ascending degeneration of the direct cerebellar tract as far as its termination in the cerebellum. The spino-thalamic fasciculus degenerates in like manner upward, as far as the thalamus, when its nuclei of origin in the posterior gray matter of the cord are destroyed. In other words, the axones when deprived of the trophic influence of their cells degenerate to their termination, that is, to the point where a new neurone begins.

Root-zones.—Anterior.—The anterior roots are formed by the massing together of the axones from the cells of the anterior gray matter of the cord. Between the gray matter and the periphery of the cord they form a dense mass of horizontal fibers, the anterior root-zones. On their way to the periphery these fibers pierce the antero-lateral commissural fasciculi, the vestibulo-spinal tract and the spino-thalamic tract. As they reach the periphery these fibers issue from the cord in separate bundles. These combine in the intervertebral foramina to form the anterior spinal roots. To this point they are accompanied by an investment of the spinal meninges. After piercing the dura they unite with the posterior roots to form the spinal nerves. The aggregations of cells within the anterior gray matter of the cord, and their prototypes in the medulla and pons, form the genetic nuclei of all of the motor nerves of the body, the function of which is to produce muscular contraction, and with the neighboring cells of the antero-lateral region of gray matter, to preside over trophic and secretory function, and to respond to sympathetic or reflex impulses from without, by motor, vasomotor, trophic or secretory activity.

A destructive lesion of the axones of these cells, either within the root-zones, anterior roots, or in the peripheral nerves, has exactly the same effect as is produced by destruction of the cells themselves, namely flaccid paralysis, loss of reflex activity, trophic disturbances, and symptoms of vasomotor paralysis (lesion of the lower motor neurone).

Root=zones.—Posterior.—The fibers of the posterior spinal roots take their origin in the spinal ganglia of these roots. They separate into several bundles before entering the cord. Soon after their entrance they divide in Y-shaped divisions and give off both ascending and descending branches. Some of these enter the gray matter either to terminate about cells that form parts of reflex arcs or cells that form relay stations in the course of the sensory tracts to the brain. Some however descend to lower levels of the cord and terminate in the spinal gray matter of these levels two or three segments below their entrance. These are the fibers that form the comma tract of Shultz (Figs. 66, 67). The majority however commence immediately to ascend, entering the columns of Burdach, and later the columns of Goll, on their way to nuclei in the medulla. Irritative lesions of the posterior roots or of the root-zones cause root pains and paresthesic sensations.

Destructive lesions produce loss or disturbance of all forms of sensation, abolition of the reflexes, disturbance of equilibrium, ataxia and trophic disturbances.

The posterior roots and root-zones are involved in posterior sclerosis (tabes dorsalis), postero-lateral sclerosis (ataxic paraplegia), and hereditary ataxia, and may be implicated in meningitis (meningo-myelitis) or tumors in or near the cord, or they may be involved by disease of the vertebræ.

The Spinal Ganglia.—On each posterior spinal root just before its junction with the anterior roots is a ganglion. In the case of the atlas and the axis they lie in the groove on the transverse processes. The first cervical sensory roots are sometimes devoid of ganglia. Below the axis they lie in the intervertebral foramina down as far as the fifth lumbar nerve. The ganglia for the five sacral nerves lie within the vertebral canal, but outside of the dural sheath. The ganglia of the coccygeal nerves alone, of all the spinal ganglia, lie within the dural sac.

All forms of sensation pass through the spinal ganglia. The sensory portions of all reflex arcs, below the medulla, pass through these ganglia. These include not only the deep muscle, tendon and bone reflexes and the superficial cutaneous and mucous membrane reflexes, but also the afferent impulses concerned in the maintenance of equilibrium and coördination, as well as sympathetic, vasomotor and visceral reflex impulses, controlling vasomotor, visceral and trophic functions.

As the cells of these ganglia are the nuclei of origin (genetic nuclei) of the posterior roots they control the nutrition of the filaments of these roots, and lesions of these ganglia are followed by degeneration through the posterior roots and root-zones as far as the terminations of the respective filaments (axones) originating in these ganglia.

Irritative lesions of these ganglia are characterized clinically by neuralgic pain in the region controlled by the ganglion affected, and often by the cutaneous phenomena of herpes zoster.

LOCALIZATION OF SPINAL LESIONS

The first step in the location of a spinal lesion is the determination as to whether the symptom-complex indicates a disseminated or a more or less limited lesion. If limited, it must be determined whether the lesion is limited vertically or horizontally. It is not necessarily a transverse lesion, although it may be so, but simply a lesion located at a certain level or levels of the cord, in contradistinction to one extending longitudinally through the cord, or one characterized by its disseminated distribution—multiple foci of disease. A lesion with vertical limitations is termed a segment lesion. A lesion limited horizontally is one that affects, longitudinally, certain conduction tracts or cell-columns in the cord. To successfully locate a spinal lesion it is necessary to keep clearly in mind the functions of the different parts of the cord and the clinical characteristics of abolition or disturbance of those functions. It must also be remembered that some parts of the cord are merely conduction paths for impulses that have their initiation and their destination in other parts of the nervous system, while other parts of the cord constitute important sources of nervous energy. The location of the vertical limits of a lesion depends upon a knowledge of the location of these spinal sources of nervous energy, and the parts of the body served by them, and the relation of the different parts of the body to the levels of the cord that correspond to the entrance of those impulses that pass through but do not originate or necessarily terminate in the cord. These latter impulses are of sensory character and afferent course, and their entrance into the cord is through the posterior roots. Only at the level corresponding to their entrance into the cord can these afferent impulses possess any value in vertical location of spinal disease. Quite otherwise however is their value in horizontal localization. Here the location of the lesion depends on symptoms of disturbance or abolition of the function of these conduction tracts, the afferent sensory tracts just referred to, and efferent motor conduction tracts from the brain through the cord.

The function of the various divisions of the cord have already been referred to in connection with the anatomy of the cord. It is now necessary to consider these functions more in detail and particularly in their bearing on spinal localization.

MOTOR FUNCTIONS OF THE CORD AND THEIR MODIFICATION BY DISEASE

The medullary and spinal motor nuclei constitute an extensive and intricate series of power stations in which is produced the nervous energy necessary for the muscular activity of the body. They are to be regarded as mere manufactories of nervous energy, the assignment and distribution of which is under the supreme control of higher cortical and sub-cortical centers in the brain.

The clearest illustration of this is afforded by the initiation and execution of a simple voluntary act.

Here the decision to act arises in the executive or administrative centers of the brain, and is transmitted to the cerebral motor cortex, whence an order is sent down through the internal capsules, crura, pons, medulla and pyramidal tracts of the cord to the particular segment of the cord where are situated the nuclei belonging to the particular muscles concerned in the contemplated act.

Lying along the entire extent of the anterior spinal gray matter are an infinite number of these subordinate power-stations in which are evolved the nervous impulses that flow out through the anterior spinal roots and the peripheral motor nerves to the muscles and cause them to contract.

Since the muscular contraction *per se*, regardless of its purpose, is produced by energy evolved in these cells of the anterior gray matter of the medulla and cord, it follows that these cells may act independently of the higher executive control and may cause muscular activity of any degree or extent. Under such circumstances, however, that muscular activity exhibits, by its lack of purpose or by its violence, the absence of systematized direction and control.

It loses therefore the character of responsible muscular activity that a voluntary act should possess, or the smoothness and moderation that even, automatic action should exhibit.

Nevertheless, even in fetal life, and to a vastly greater degree from the moment of birth on through the subsequent years, these subordinate power stations of the medulla and cord are called upon to answer innumerable demands of a more or less automatic nature, in which the higher centers are not directly or immediately interested. In fact to require those higher centers to consume their time with personal attention to those details would be an economic mistake and leave them little time for any other duties.

Clinical experience, however, teaches us that, while the performance of these perfunctory acts is relegated to these subordinate power stations of the medulla and cord, they are nevertheless not left entirely without supervision of the manner in which they perform those acts. Such supervision probably devolves upon subcortical centers in the lenticular nucleus, corpus striatum, thalamus and cerebellum.

As long as the cortical tracts to the cord are intact voluntary initiation of muscular activity is possible, and as long as the subcortical centers retain their communication with the spinal nuclei, both voluntary and automatic acts may be properly performed. Whenever, on the other hand, the higher centers are cut off from those nuclei immediate disturbance of normal muscular function results.

Thus if some lesion occurs at any point in the pyramidal system, voluntary control of the activity of all the spinal nuclei to which the particular pyramidal fasciculi affected run, will be lost or interfered with, normal muscle tone is replaced by increased tonicity—muscular spasm—and deep reflex activity is increased. If the lesion lies in the cord proper, that is, below the pyramidal crossing in the medulla, the muscles affected will be those that are on the same side as, and below, the lesion.

The pyramidal system is often termed the upper motor segment. The lower motor segment commences in the anterior spinal nuclei and is projected by the axones of these cells through the anterior roots and peripheral motor nerves to the muscles.

Lesions of the upper motor segment, as will be noted by the above mentioned symptoms, do not cause paralysis in the true sense of the term, but a loss or impairment of volitional control over muscular action, while the coincident impairment of normal inhibition of automatic muscular activity results in muscular spasm and exaggeration of reflex activity. In other words there is an increase of muscular activity, but it is of an irresponsible character. Increased tonicity of the muscles or muscular spasm is characterized by involuntary resistance to active voluntary movement, or by involuntary resistance to passive movement of the part affected, or by both active and passive spasm. Spasmodic muscular twitchings may occur in any spinal lesion involving the pyramidal tracts. These involuntary movements may take the form of clonic tremor-like oscillations of an extremity or non-rythmical more or less violent extension or flexion movements. In addition to exaggeration of the normally present deep reflexes, there often appear certain reflex phenomena never seen except in upper motor segment disturbance. These will be described later under the heading of abnormal reflexes. Whereas the deep reflexes are usually exaggerated in lesions of this part of the motor system, the opposite is the case with the superficial normal reflexes. They are lost in upper motor segment lesions.

As the symptoms of upper segment lesions have the same characteristics regardless of where they may happen to be located in that segment, they are not, *per se*, of value in segment localization of spinal cord disease. In order to locate accurately the particular level of the cord at which the pyramidal tracts are involved other and localizing symptoms must be associated with the spastic phenomena.

If such symptoms are not present reliance must be placed on the history of the manner of onset and development of the spastic phenomena and their correspondence to known types of spinal-cord disease.

A very different clinical picture results when there is a lesion in the lower motor segment. This segment, as we have seen, commences in the anterior spinal nuclei, whose axones form the anterior spinal roots and the motor fibers of the peripheral nerves. These anterior spinal nuclei furnish the impulses that produce muscular contraction, irrespective of whether that contraction is volitional or not. For the occurrence of muscular contraction as the result of nervous energy it is essential that this lower motor segment remains intact. Any destructive lesion within the fountain head of this nervous energy—the anterior spinal nuclei—or any break in its conduction path to the muscles, results in absolute paralysis. In other words, the paralyzed muscles completely lose their tonicity, and are found in a state of complete relaxation or flaccidity. We have seen that this lower motor segment forms the motor portion of all the spinal reflexes. A lesion in this segment, therefore, abolishes the reflexes. Moreover certain of these anterior spinal nuclei control that automatic sympathetic mechanism comprehended in the term vasomotor function. When these nuclei or their axones are destroyed vasomotor paralysis results. It is evident clinically by a reduction in the temperature of the part of the body affected and often, although not invariably, by cyanosis of the part. Other of these nuclei seem to have some influence over the nutrition of the muscles. Therefore, trophic disturbance in the muscles is a conspicuous feature of a lesion in this lower motor segment, and the degeneration and consequent shrinking in the affected muscles is too rapid and too extreme to be explained as merely the result of disuse incident to the paralysis. Moreover in muscles paralyzed by such a lesion, an alteration occurs in the manner in which they respond to the stimulation of faradic and galvanic electricity, an alteration never seen in paralysis caused by lesions in the upper motor segment. This is called the reaction of degeneration. When complete it consists of a loss of response of the muscle to the faradic current, an increased excitability to the galvanic current, in that the muscle responds to a lesser intensity of current than will produce contraction in normal muscle, a change in the quality of that contraction, in that instead of being instantaneous and lively, it becomes sluggish and "worm-like," and an alteration in the muscle's manner of response to the negative and positive poles.

Normally the "making of the current"—the closure of the circuit—with the negative electrode on the muscle—cathodal closure contraction—produces more marked contraction than when the positive electrode is similarly employed—anodal closure contraction.

When, however, the complete reaction of degeneration is present exactly the reverse is the case, the anodal closure contraction being greater than the cathodal closure contraction.*

Various stages leading up to the complete R. D., as it is called, naturally modify the condition as above described. Thus, in the early stages, muscular response to both faradism and galvanism may be diminished. Moreover in partial lesions the reaction of degeneration may never reach its complete stage. Where the lesion is severe the complete R. D. may be obtained in from two to three weeks after the onset of the lesion.

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* The above features of the reaction of degeneration are all that it is essential to employ from a practical clinical standpoint. Therefore the polar opening muscle response and the nerve response are here omitted.

The normal response of muscle is expressed by the formula:

$$\text{Ca.Cl.C.} > \text{An.Cl.C.}$$

The R. D. response is indicated by the formula

$$\text{An.Cl.C.} > \text{Ca.Cl.C.}$$

In partial R. D. the formula is sometimes

$$\text{An.Cl.C.} = \text{Ca.Cl.C.}$$

Briefly expressed, the features of lower segment paralysis are:

Flaccid paralysis
Loss of reflexes
Muscular atrophy
Vasomotor paralysis
(R. D.)

The characteristics, therefore, of a lesion of the anterior spinal ganglia, or of their axones in the anterior spinal roots, are well defined, and because these characteristic symptoms correspond exactly to the level of the cord in which the lesion causing them is situated, we shall find that these symptoms are of very great value in segment localization of spinal cord disease.

In slow degenerative lesions of the anterior spinal nuclei the phenomenon known as fibrillary muscular twitching is often seen. This consists of a peculiar more or less fleeting and recurring tremor of individual muscle bundles, somewhat similar to that seen on exposure of a previously covered part of the body to cold.

While lesions of the upper or lower motor segments are the most frequently met with, there are other motor disturbances seen in spinal disease. These are disturbances of coördination and of equilibrium. They are usually due to lesions, not in the motor but in the sensory tracts of the cord or in the posterior sensory roots.

These are the tracts that convey to the coördinating centers knowledge of the position of the body and its parts, and their relation to their environment. They are conduction pathways through the cord on their way to the medullary nuclei and the cerebellum.

Similar disturbances should occur in lesions involving the centrifugal vestibulo-spinal tracts, but little is known clinically in regard to involvement of these particular pathways.

Disease in the spinal cord may be chiefly or wholly limited to one particular fasciculus, and extend throughout this particular bundle of fibers. Such affections are called system diseases. Often there is simultaneous involvement of some other system of fibers and such affections are designated combined system diseases.

The term is one mainly of convenience in designating affections that seem prone to follow certain tracts or systems, in contradistinction to scattered foci of disease or to lesions limited to a particular segment of the cord. These affections are usually progressive sclerotic processes with ascending or descending degeneration through the conduction tracts of the cord. Among the typical instances of these system diseases are primary lateral sclerosis, tabes dorsalis, hereditary spastic paraplegia, hereditary cerebellospinal ataxia, and among the combined system diseases postero-lateral sclerosis and amyotrophic lateral sclerosis are representative. Some of these affections are familiar, in that they show a tendency to affect several members of one family, or to extend through several generations.

These diseases are located by the characteristic clinical signs indicative of disturbance of the functions of the particular fasciculi involved. These clinical signs may, in other words, be said to be classified or systematized, in contradistinction to the heterogeneous symptoms of scattered lesions in the cord, or in contrast to the combination of symptoms seen in vertical or segment lesions.

Nevertheless the term is an elastic one and cannot be too rigidly construed, for some of these diseases present in their earlier stages symptoms indicative of well-defined limitation to system tracts, but eventually show evidence of wide spread involvement throughout the nervous system.

The symptoms of these affections will be evident from what has been said of the functions of the various tracts of the cord, and many of them have been assigned to other colleagues for detailed consideration.

The accompanying table by Church,* somewhat altered by the writer, gives a concise idea of the classification of the various diseases of the cord.

Organic diseases of the cord	Indiscriminate lesions	Hemorrhage Myelitis Softening Insular sclerosis Tumors Traumatic injuries Compression from bone diseases in tumors		
	System disease	Disease of single tracts	Anterior horns	Acute poliomyelitis Progressive spinal muscular atrophy
			Lateral pyramidal	Primary lateral sclerosis Hereditary spastic paraplegia Lathyrism
			Posterior columns	Tabes dorsalis Ergotism
		Combined disease	Anterior horns and lateral pyramidal tracts	Amyotrophic lateral sclerosis
			Lateral and posterior columns	Ataxic paraplegia Cerebellospinal ataxia Pellagra Inflammatory or traumatic transverse lesions of the cord

In the description of the anatomy and physiology of the cord it will be recalled that groups of cells in the anterior gray matter of the cord controlled the various muscles of the body, that the axones of these cells forming in bundles leave the cord and soon uniting become

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* Church and Peterson, "Nervous and Mental Diseases." Saunders (1911), p. 356.

the anterior motor roots, that, after joining the corresponding posterior sensory root, they form the mixed sensory-motor spinal roots, and that these unite with adjacent similar roots to form plexuses from which the peripheral nerves are derived. It is evident therefore that every peripheral nerve may contain fibers from more than one level of the cord, and it has been demonstrated that a large proportion of the muscles of the body are supplied by fibers from more than one level of the cord. It follows then that a lesion strictly limited to a single segment of the cord does not usually result in total paralysis of any one muscle.

If a single muscle is completely paralyzed it points usually to a lesion in the peripheral nerve supplying that muscle rather than to a lesion within the cord.

Nevertheless there are exceptions to this rule and often the history of the manner of onset and character of the course of the paralysis, and its association with other symptoms, must be carefully considered before conclusive localization of the lesion can be made.

Although no one muscle or muscle-group corresponds to any single segment of the cord, nevertheless a knowledge of the approximate localization of the spinal centers for the various motor functions of the body is absolutely essential in the focal diagnosis of spinal disease, for on this localization and the associated segmental sensory involvement the vertical localization of spinal lesions depends.

In the table on page 585 will be found the approximate segmental location of some of the simple movements of the various parts of the body, movements of practical utility in clinical tests of motor function, and all of which may be easily carried out, even in young children, especially if the incentive of play is introduced in the tests.*

A partial transverse lesion of the cord produces a flaccid paralysis of the muscles innervated by that region, while all muscles below the level of the lesion will show paralysis of the spastic type (paraplegia) from involvement of the pyramidal fasciculi at the point where these fasciculi traverse the lesion. A total transverse lesion of the cord results in a complete flaccid paralysis of all muscles at and below the lesion, with abolition of all reflex activity. If, however, the transverse lesion is severe but not entirely destructive, after a few days or weeks conduction through some of the pathways may be partially restored, and the clinical picture of a complete transverse spinal lesion is considerably modified. Thus after severe pressure on a transverse section of the cord, and particularly as the sequela of a transverse myelitis, we may eventually have the symptoms of spastic paraplegia, described as characteristic of a lesion of the pyramidal tracts alone.

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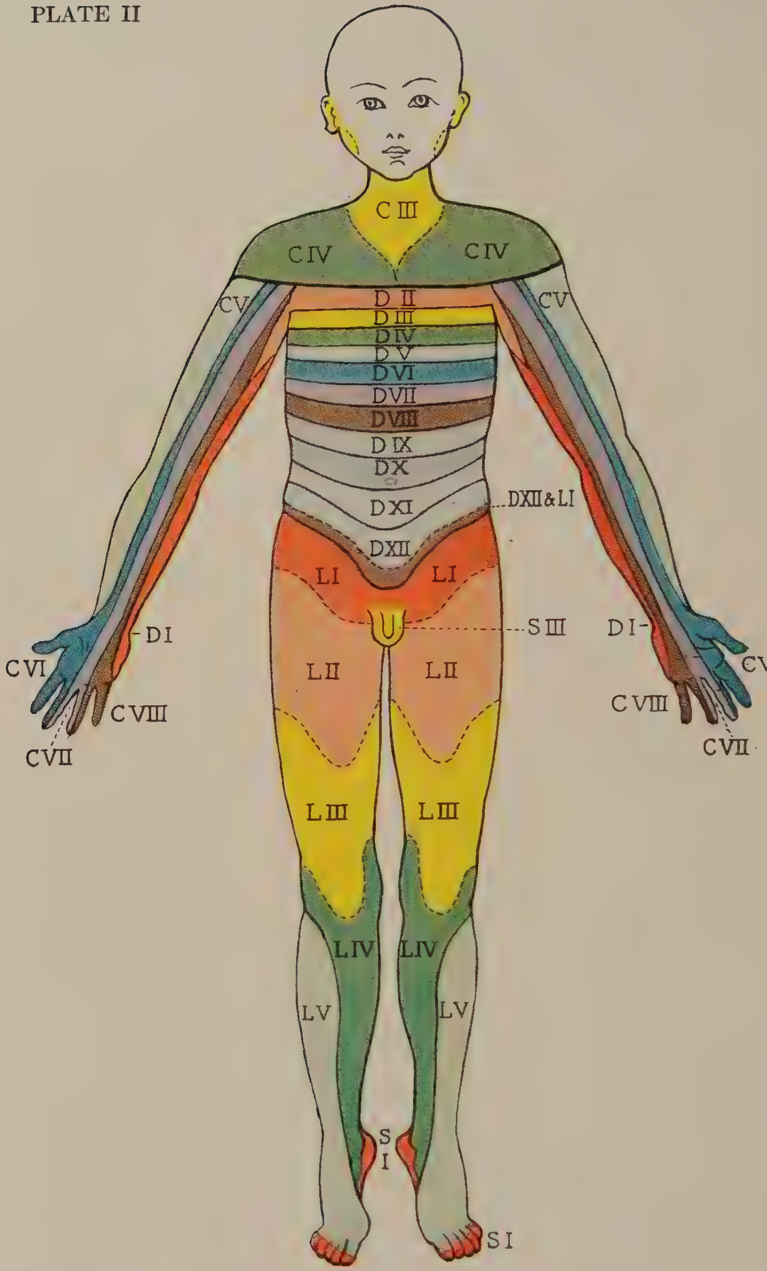
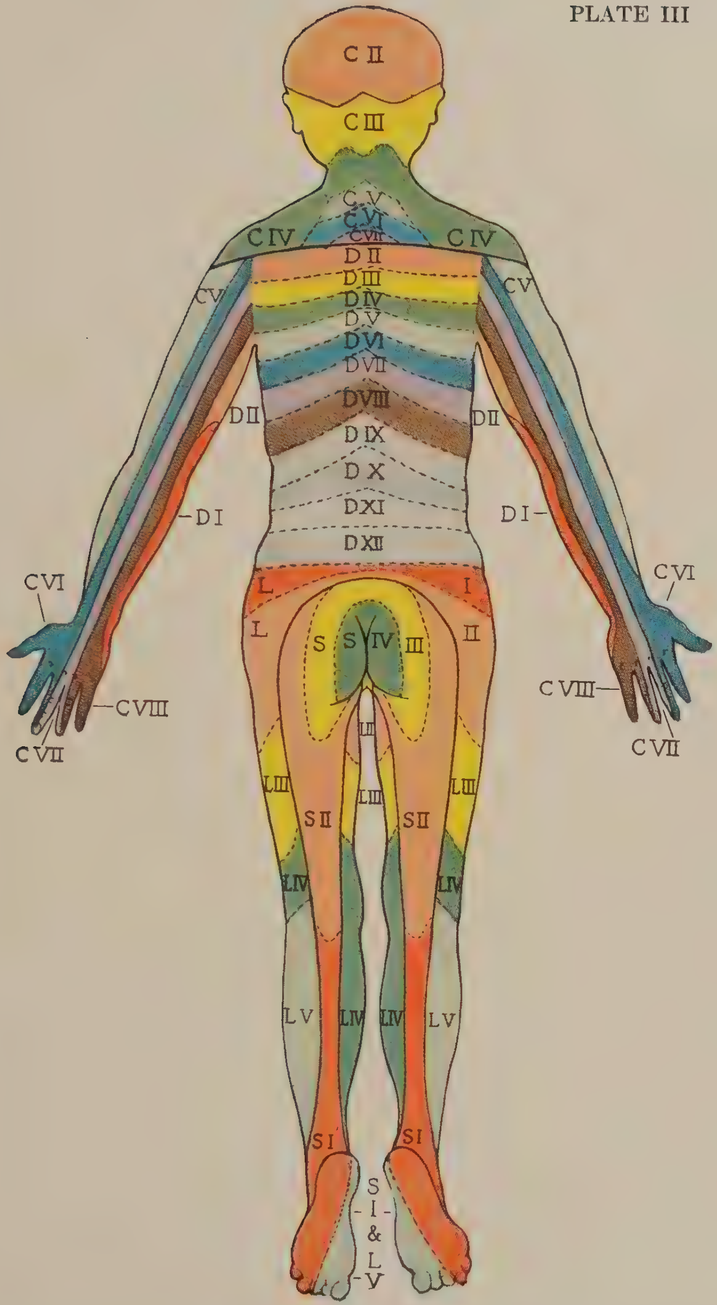
* It is not possible to enumerate here the methods employed to secure the coöperation of the child in carrying out these tests. Much must depend therefore on the patience and ingenuity of the examiner. Many suggestions along this line may be obtained in the excellent little book on "Educational Gymnastic Play for Little Folks" by Johnson and Colby, Educational Publishing Co., Chicago. These are the methods employed for many years at our clinic at the Boston Children's Hospital, in the treatment of paralytic children.

PLATE II



Cutaneous sensory segment distribution of the spinal cord. The segment areas are indicated, in each division of the cord, by the colors of the spectrum (Renz), with the addition of brown for the eighth cervical and the eighth dorsal. The ninth, tenth, eleventh, and twelfth dorsal are left uncolored, except in the lower area of the twelfth, just above Poupart's ligament, where the supply is very variable, but where LI probably dominates in the average case. Dotted boundaries indicate variable areas, areas that not merely overlap, but that appear to vary considerably in different instances. Again it must be emphasized that these boundaries are to be used only as guides to the maximum segment distribution in each area. Some allowance must be made for supply from the segment above and below, except between CIV and DII, and LI, II, III, IV and SI, II, III, where overlapping does not occur.

PLATE III



Cutaneous sensory segment distribution of the spinal cord. The ramus posterior of CI does not take part in the sensory innervation of the scalp. Its branches are exclusively muscular (rectus capitis posterior, major and minor, and obliquus superior and inferior). It is an exception to the corresponding posterior divisions of the other spinal nerves.

Over the inner half of the sole of the foot LV and SI share the supply, but to avoid confusion this territory is left to LV.

Sacral V, with supply about the anus, is not visible in this diagram.

Table 1.—Approximate Segment Localization of Certain Voluntary Muscle-functions

Forward and backward movement of the head	C 1–2
Lateral flexion and rotation of the head	C 1–2
Raising of the shoulders	C 2–4
Outward rotation and abduction of arm	C 4–6
Flexion and supination of the forearm	C 5–6
Extension of the forearm	C 6–7
Extension of the wrist (radial side)	C 6–7
Extension of the wrist (ulnar side)	C 7–8
Flexion of the wrist (radial side)	C 6–7
Flexion of the wrist (ulnar side)	C 7–8
Extension of the fingers and thumb	C 6–7
Abduction of the thumb	C 6–7
Inward rotation of the arm	C 7–8
Supination of the forearm	C 5–6
Pronation of the forearm	C 6–8
Apposition of the thumb and fingers	C 6–8
Adduction of the thumb	C 8
Abduction and adduction of the fingers (except the thumb)	C 8–D 1
Flexion of the thumb	C 6–7
Flexion of the fingers	C 7–D 1
Anterior, lateral, and posterior movements of spine	D 2–12
Fixation of abdominal muscles (as in raising of head, etc.)	D 5–L 1
Raising of the body from the recumbent position	L 1–4
Flexion of the thigh	L 1–4
Crossing of the thighs	L 2–4
Extension of the legs	L 2–4
Inward rotation and adduction of the thigh	L 2–4
Flexion of the legs	L 4–5
Extension (and inward deviation) of the foot	L 4
Extension (and outward deviation) of the foot	L 5–S 1
Extension of the toes	L 5–S 1
Flexion of the foot (standing on the toes)	L 5–S 1
Flexion of the toes	L 5–S 2
Outward rotation and extension of the thighs	L 4–S 2

The motor symptoms of circumscribed lesions of a cord segment will be evident from what has already been said in connection with the various tracts and ordinary motor nuclei of the cord. The effects of destruction of one-half of the cord will be described later—Brown Séquard paralysis. The involvement of special organic motor centers is also considered later.

SENSORY FUNCTIONS AND THEIR DISTURBANCE BY SPINAL CORD DISEASE

Different forms of sensation do not all pursue the same pathways through the cord on their way to the brain. As was seen in the consideration of the different fasciculi and their functions, most of the transmission of tactile and all of the transmission of muscle sense is via the posterior columns.

Pain and temperature sensations, on the other hand, are sent through the gray matter for a short distance, cross in the anterior gray commissure in front of the central canal, and thence enter the antero-lateral columns.

This results in a dissociation of sensory disturbance in certain spinal lesions, a separation of the different forms of sensation in the way they are influenced by localized lesions of the cord.

Since all of these forms of sensory perception pass through the posterior roots, it follows that root lesions usually implicate all forms of sensation to some degree, although even here, as in lesions of the peripheral nerves, some forms of sensation may be earlier or more severely affected than other forms.

No given area of skin is supplied with sensation solely by any single posterior root, but every portion of the cutaneous surface is supplied by at least two posterior roots. This fact, first demonstrated by Sherrington (Transactions of the Royal Philosophical Society, London, clxxxiv) (1893) is often referred to as Sherrington's Law.

In other words, the sensory filaments from the different posterior roots overlap in their cutaneous distribution. Destruction of a single posterior root therefore does not result in loss of sensation in the area of skin in which its filaments originate. There is some variation in the extent to which this overlapping occurs in different cutaneous surfaces of the body. It is most extensive on the palms of the hands and soles of the feet, and is most complete for the tactile sense. In the furtherance of clinical localization of spinal cord lesions the endeavor has been made by many investigators to represent graphically the exact areas of skin that correspond to individual spinal roots. The results unfortunately have been conflicting, especially for the cutaneous areas supposed to correspond to the cord segments below the twelfth dorsal. In fact for this portion of the body surface no two charts are alike and some do not even approximate similarity. Undoubtedly there exists some degree of variation in different individuals in the sensory distribution over the body surface. Nevertheless an approximate localization is of very great practical value, and often it is from this sensory localization alone that we are able to determine the probable site of the lesion. The accompanying diagrams (Plates II, III) present the approximate cutaneous sensory involvement in the lesions of the various levels of the cord.*

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* In using these diagrams for clinical segment localization it must be kept in mind that there is everywhere an overlapping of contiguous areas, except between C 4 and D 2 in the upper thoracic region, and between L 1–4 and S 1–4 on the back of the thigh and buttocks.

It is well to remember that the segment lesion in any clinical case probably lies somewhat higher than would seem to be indicated by the zone of absolute anesthesia.

The areas here given are those of the maximum segmental supply for the respective levels of the cord. In each area, except those above refered to, allowance must be made for some overlapping of the segment above and below, especially in the thoracic and abdominal region.

These areas of segmental supply here given are the result of extensive study of the anatomical work and clinical reports of numerous investigators. It is believed that they will be found to accord with the average clinical experience. As Paterson has shown, there is considerable variation in the segmental origin of nerve trunks and this with variation in peripheral anastomosis will account for the variations

A complete transverse lesion of the cord produces complete sensory loss at and below the level of the segmental area involved. Just above the level of absolute sensory loss—anesthesia, analgesia, thermoanesthesia—will often be found a band of hypersensitiveness, hyperesthesia, etc., corresponding to those sensory conduction paths in the sensory roots and in the cord, that touch upon and are irritated but are not destroyed by the lesion. In such a hypersensitive area not only may all forms of sensation be perceived with increased intensity but they may also be perverted—paresthesia—and be interpreted by the patient as prickling, tingling, crawling, burning sensations, numbness, etc.

The patient may also describe a sensation of constriction, corresponding to this area, and felt as a girdle about the body or as tight gloves on the hands, etc. This is due to irritation of the posterior nerve roots, and may not only be complained of under the above mentioned circumstances but whenever the posterior roots are irritated.

Pain may be also present in this hypersensitive area, and when so, it also is caused by irritation of the posterior roots. It may be of severe lancinating character, and its location corresponds to the peripheral distribution of the posterior roots affected. A lesion within the cord and not involving the posterior roots does not cause pain of this character or this distribution. In fact such lesions are often unattended by spontaneous pain. Circumscribed lesions of the cord, especially such as are limited to the gray matter about the central canal, may produce dissociated sensory disturbance. In such cases perception for temperature may be entirely lost and pain perception may be partially involved or entirely lost, while tactile perception is unaffected. This is seen in syringomyelia and occasionally in limited hemorrhages within the central gray matter, or in central tumors. While dissociated sensory symptoms usually point to disease of the central gray matter, they may also occur as a result of implication of the antero-lateral columns of the white substance of the cord, either from disease within these tracts or by pressure from without.

In such cases simultaneous involvement of neighboring white fasciculi is likely to occur, especially the several adjacent cerebellar tracts or the vestibulo-spinal fasciculi, with resulting disturbance of coördination and equilibrium. If the lesion is within the gray matter it is not likely to remain long limited to the region about the central

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in segment diagrams, and for variation in certain clinical cases. Thus the variation in the territory ascribed to the 12th. thoracic and the 1st. lumbar segment is easily explained be the fact that Paterson found the 12th. entering into the formation of the lumbo-sacral cord in one case, and in four out of seven instances L 1 was the first root of the anterior crural nerve, and he also states: "In the plexuses which I have examined L 1 has frequently been concerned in the constitution of the genito-crural and external cutaneous nerves." Moreover Hirschfeld and Leveillé indicate L 1 as the source of the chief root of the genitocrural nerve. From these facts it will be apparent that any diagram to conform to all cases cannot be constructed.

portion but extends to the anterior region and is eventually associated with muscular atrophy and paralysis. Such association locates the lesion within the gray matter rather than in the lateral columns of the cord. In disease or injury involving only the posterior columns tactile and muscle sense are seriously impaired, while pain and temperature sensations may be normally perceived, unless, as is usually the case, the posterior roots are also implicated.

It should be remembered that in incomplete transverse lesions conduction for the more delicate forms of sensation may be broken while painful stimuli may still be transmitted. In incomplete transverse lesions sensory stimuli applied to the anesthetic area below the level of the lesion are said by Starr to be sometimes felt in the zone of hyperesthesia just above the level of the anesthesia. I have not happened to see this myself.

A not uncommon occurrence in tabes is a delay in the perception of sensory stimuli, the recognition of the stimulus occurring several seconds after it is applied. This simply means delayed transmission through partially diseased conduction tracts.

A common accompaniment of the sensory disturbances met with in posterior root disease is trophic disturbance in the territory supplied by the diseased root. The reason for this occurrence can only be explained on the supposition that the trophic centers of the anterior gray matter of the cord are in some way dependent on unimpaired sensory connections with the periphery for the maintenance of nutrition and the repair of damaged tissue. Trophic ulcers and bedsores are among the most common and troublesome sequelæ of spinal cord disease.

SPINAL REFLEXES AND THEIR BEHAVIOR IN SPINAL DISEASE

As has already been mentioned in connection with the preceding outline of the anatomic structure of the cord, many of the nuclear cells of the gray matter of the cord and many of the white fibers originating in or terminating about these nuclei are parts of reflex mechanisms, the greater number of which are purely automatic and largely independent of the higher volitional centers.

Some of these mechanisms are of highly complex and specialized character. Of these only a part can be regarded as exclusively or peculiarly spinal. As we shall see, many of these reflex mechanisms have only part of their structure within the spinal cord proper.

On the other hand, there are innumerable relatively simple reflex phenomena that may be regarded, for all practical clinical purposes, as being of exclusively spinal origin.

DEEP TENDON AND BONE REFLEXES

The simplest of these are the phenomena induced by tapping a muscle or its tendon, or the adjacent bone, a procedure that results in a contracting of the respective muscle or muscles and consequent movement of the part supplied by the muscles. In this instance the occurrence is to be explained by the production, by means of the peripheral stimulation of the blow on the tendon or bone, of a centripetal sensory impulse that travels via the sensory filaments of the peripheral nerves to and through the spinal ganglia and posterior nerve roots to the receptive nuclei in the gray matter of the posterior portion of the cord, whence the impulse is relayed to the motor cells of the anterior horns and there excites a motor impulse that proceeds through the anterior roots and motor filaments of the peripheral nerves and causes contraction of the respective muscle. Reference to the diagram (Fig. 69) will indicate the comparative simplicity of this type of reflex.

This type of reflex is called a deep reflex or a tendon or bone reflex in contradistinction to another common type of reflex to be described later.

There are many of these deep reflexes, and all may be utilized in the diagnosis of diseases of the nervous system, but only a few are essential and commonly employed. The table on p. 593 indicates those of common utility, together with the segment of the cord through which the reflex impulse passes.

The mechanism through which the reflex action is brought to pass is termed a reflex arc. In the simple type of reflex just described, it consists essentially of two parts, a centripetal sensory and a centrifugal motor pathway.

A break in any part of this arc abolishes the reflex. A break in the sensory pathway is likely to be associated with sensory symptoms of one form or another. A break in the motor pathway is likely to be associated with motor paralysis. In the latter case it is the lower motor neurone that is involved and the paralysis, if present, has all of the characteristics described as peculiar to lower motor segment lesions.

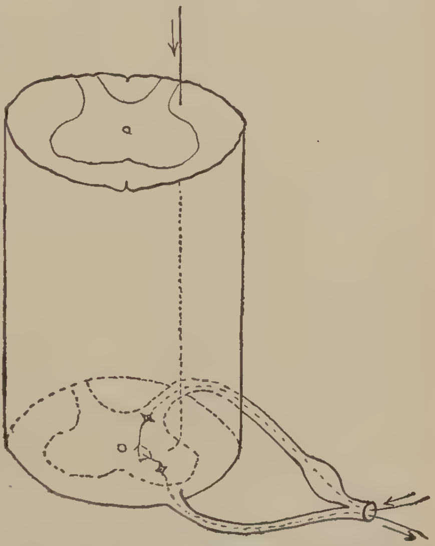

Fig. 69.—Diagrammatic representation of a reflex arc, showing the centripetal sensory path, the centrifugal motor path, the synopses, and the inhibitory path from the brain.

Detailed description of the method of eliciting the various normal tendon and bone reflexes is hardly practicable within the limits assigned to this chapter, but a few general principles must be kept in mind if success is to be achieved, and if the results obtained are to be properly interpreted. This is particularly true in respect to the correct interpretation of the results obtained in childhood. During many years of teaching neurology, and more especially in the teaching of methods of examination in the nervous diseases of childhood to post-graduate students, I have been impressed with the necessity of intensive training in all the details of technique, and in none more so than in the proper eliciting and estimation of the value of reflex phenomena.

No matter how experienced the examiner may be in the case of adults, he is sure to be perplexed in the examination of children if he gives the same value to the apparent results obtained that he has been accustomed to rely upon in older patients.

There are, however, a few general principles that must be observed in all cases, either adults or children.

In the first place the utmost relaxation of the muscles to be tested must first be obtained. This is sometimes extremely difficult, particularly in the upper extremities. With the best of will on the patient's part, there often ensues, the moment the eliciting of the reflex is attempted, a subconscious rigidity of the extremity which it is beyond the patient's power to overcome. The more he is urged to relax, and the more he attempts to do so, the more incapable he becomes. After a few attempts have proved unsuccessful, it is best to forego the attempt for a time and take up some other part of the examination. Thus, for instance, in the attempt to test the knee-jerks the patient should be placed on a table, if possible in a sitting position, and while their attention is diverted by the examination of some other part of the body, opportunity is sooner or later afforded for a quick and unexpected tap on the patella tendon and the reflex is easily and accurately obtained. In children lack of relaxation is usually a much less disturbing factor than is the interference occasioned by voluntary restless movement.

In both adults and children it is better not to proceed abruptly with the testing of the reflexes, but by diverting conversation of an irrelevant character, to place the patient entirely at his ease.

In the second place the blow on the tendon or bone must be quick and clean, not heavy or awkward or sufficiently intense to cause pain. Many individuals are quite sensitive over muscles, tendons and bones, and this is particularly the case in children, and forcible compression of the poorly developed muscle or tendon between the pleximeter and the bone may cause so much discomfort that all further attempts to elicit the reflex are frustrated.

In the third place it should be remembered that there is no arbitrary limit of minimum or maximum intensity of any reflex. The limits are elastic, and the estimation of the character of any reflex must be qualified by the existence or non-existence of other signs, and by the proper valuation of those associated symptoms when they are present.

Furthermore no deep normal reflex should be declared absent until the method of reinforcement described by Jendrassik has been employed. This consists of the production of voluntary muscular effort in some other part of the body simultaneously with the blow on the tendon of the muscle that is being tested. There are many ways of doing this. In the case of the lower extremities a simultaneous pull by the patient on his clasped hands in the eliciting of the knee-jerks will often bring to the front an otherwise supposedly absent reflex, or gripping the examiner's hand may suffice to reinforce the reflex impulse, or in the case of the ankle-jerk—Achilles—gripping the back of a chair on which the patient is kneeling is frequently employed. With patience even quite young children may be induced to thus coöperate with the examiner, especially if the procedure is given the outer semblance of play.

In examining the deep reflexes in childhood one important fact is especially to be emphasized and this is that these reflexes are easily exhausted. The reflex arc seems to quickly tire of transmitting impulses. The very fact that examiners do not always observe in the examination of children, the rules above mentioned as necessary even in the examination of adults, and the resulting perplexity as to the meaning of the response obtained, leads to the repetition of the test several times in quick succession. The ambiguity of the earlier response is increased by the eventual abolition of all response and the apparent absence of the reflex. I have frequently seen the examiner bewildered by this natural result of defective technique. A period of rest must be allowed the reflex arc before further conclusions can be drawn. It will usually be found that a brief rest will restore the transmission of the reflex impulse.

Even in the simplest reflex arc the centripetal sensory impulse has not only to meet the resistance of the sensory filaments *per se*, but it must overcome at least one, and probably two, synapses, before it reaches the anterior motor nuclei. These synapses are the points of connection between the terminal filaments of one neurone and the cell body or commencement of the next neurone of the series through which any nervous impulse may have to pass. Since these synapses form a break in the continuity of the transmission lines, a certain resistance is there encountered, and a certain expenditure of energy, over and above that required for mere transmission through an unbroken circuit, is presupposed to be necessary if the impulse is to jump this break. It is probable that in childhood a frequent repetition of the stimulus so fatigues the sensory filaments that, while the sensory impulse is transmitted as a sensation through the sensory pathways, it is not carried with sufficient energy to cross the break into the motor portion of the reflex mechanism. I regard this as more probable than fatigue of the motor neurone, since the actual expenditure of muscular energy in such reflex acts is infinitesimal compared with the extraordinary capacity of children to endure unremitting voluntary muscular activity.

Whatever explanation may be accepted, however, great caution must be observed in declaring a reflex to be absent in childhood on account of this factor of fatigue, as well as the other factors mentioned as often interfering with the eliciting of reflexes. In infants and in very young fat children it is often very difficult, if not impossible, to bring out a tendon reflex because of the smallness of the tendon and the thick overlying cushion of subcutaneous fat. In such instances it is better to abandon the attempt than to be led into a diagnostic error by the easy assumption that the reflex is non-existent.

The deep tendon and bone reflexes are exaggerated in all lesions of the pyramidal system—upper motor segment lesions—and occasionally in irritative lesions of the anterior or posterior nerve roots. They are sometimes very active in purely functional disturbances, as in neurasthenia, psychasthenia and hysteria. They are diminished or abolished in lesions anywhere in the reflex arc. Rarely certain of them appear to be absent where no cause for that absence can be discovered.

SUPERFICIAL CUTANEOUS REFLEXES

Less simple and less easily understood than the reflexes just referred to are those excited by irritation of cutaneous surfaces. There are innumerable such reflexes but a few only are of common utility. Reference to the table of reflexes will indicate the chief among these and the cord level to which they correspond. This level does not however represent more than a small portion of their reflex arc. These reflexes differ materially from those just described in the manner in which they are affected by organic disease. Whereas the deep reflexes are exaggerated by lesions in the pyramidal system, these reflexes are abolished by such lesions, and they are never increased by organic disease. An exception to this rule should perhaps be made in the case of the cutaneous pupillary reflex (dilation of the pupil on irritation of the skin of the cervical region). I have seen this reflex apparently increased in meningeal irritation. The other cutaneous reflexes are sometimes unusually active in functional nervous disturbances. Their activity varies widely within normal limits, and their absence is never to be interpreted, in itself alone, as indicative of disease, but only as of confirmative value in association with other and more significant phenomena. Any break in the reflex arc naturally abolishes these reflexes, as it does all others under like circumstances, but the loss of any of these reflexes cannot be given the same value in segment localization of spinal lesions that is accorded the loss of one or more of the deep reflexes. The reason for this is obvious from what has been said, for it will be noted that a lesion in either the upper or the lower motor neurone destroys or interferes with these reflexes, therefore their loss does not necessarily depend on a lesion of any one segment of the cord. There is good reason for the hypothesis that these reflexes may be of cerebral origin, or at least that their reflex arc is much more complex than that of the deep reflexes, and that a portion of that arc lies within the pyramidal system.*

The lesion in the pyramidal tracts abolishing these reflexes is not necessarily a destructive lesion, since mere interference with conduc-

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* This subject is considered by Fairbanks in an article on the contralateral plantar reflex, Review of Neurology and Psychiatry, Edinburgh (1911), p. 225.

TABLE 2

DEEP TENDON AND BONE, AND SUPERFICIAL CUTANEOUS REFLEXES*

Tendon-bone reflexes	Cutaneous reflexes	Produced by	Resulting in	Spinal segment, localization of arc
	Pupillary	Irritation of the skin of the neck	Dilation of the pupil	C, 4–D, 1
	Scapular	Irritation of the skin over scapula	Contraction of scapular muscles	C, 5–D, 1
Biceps		Tapping the tendon	Flexion of forearm	C, 5–C, 6
Radius		Tapping styloid process of radius	Supination of forearm	C, 5–C, 6
Triceps		Tapping the tendon	Extension of forearm	C, 6–C, 7
Scapulo-humeral		Tapping the inner lower edge of scapula	Adduction of arm	C, 6–C, 7
Carpal extensor		Tapping extensor tendons of wrist	Extension of hand	C, 6–C, 8
Carpal flexor		Tapping flexor tendons of wrist	Flexion of hand	C, 7–C, 8
	Palmar	Irritation of palm of hand	Flexion of fingers	C, 8–D, 1
	Epigastric	Irritation over lower ribs	Indrawing of the epigastrium	D, 7–D, 9
	Upper abdominal	Irritation of skin of the upper abdomen	Indrawing in the hypochondrium	D, 8–D, 9
	Lower abdominal	Irritation of skin of the lower abdomen	Indrawing in the lower abdomen	D, 10–D, 12
	Cremaster	Irritation of skin in femoral region	Elevation of testis	L, 1–L, 2
Patellar (knee-jerk)		Tapping patella tendon	Extension of leg	L, 4
	Gluteal	Irritation of skin over the glutei	Contraction of the gluteal muscles	L, 4–L, 5
Achilles (ankle-jerk)		Tapping Achilles tendon	Plantar flexion of foot	L, 5–S, 1
	Plantar	Irritation along the sole of foot	Flexion of toes	S, 1–2
	Anal	Prickling of skin of perineum or introduction of finger into the rectum	Contraction of the sphincter ani	S, 5

* Arranged from the works of Wichman Flatau and Bolk. Wichman, "Die Rückenmarksnerven und ihre Segmentbezuge." Berlin (1900).

Flatau: Die motorischepensible und Reflex-Segmentierung im Rückenmark, Handbuch der Neurologie Lewandowsky, Band I (ii) p. 623.

Bolk: Morphol. Tahrb. xxv (1897) p. 465. Ibidem xxvi (1898) p. 91.

tion through these tracts may for the time being abolish the reflex transmission.

The superficial reflexes are easily elicited and those included in the table are practically always present. Rarely one or more may be absent for no demonstrable reason. In the child voluntary movement often interferes with success in obtaining the plantar especially, and the abdominal and epigastric cannot be elicited while a child is crying. In testing the cremaster in infants and young children care should be taken to determine beforehand that the testicles have descended into the scrotum, otherwise this reflex might be construed as being absent in cases of cryptorchism.

In testing the plantar reflex not too pointed an instrument should be employed for the plantar surface is usually extremely sensitive.

ABNORMAL REFLEXES

Under this designation are included certain reflex phenomena never seen except under abnormal conditions. They are all indicative of lesions of, or interference with conduction in, the upper motor neurone (pyramidal system).

Babinski Reflex.—The most valuable of these reflexes from a practical clinical standpoint is that known as Babinski's reflex. It is also one of the most interesting since it is closely associated with one of the normal superficial reflexes. Normally when the sole of the foot is stroked, from the heel forward to the ball of the foot, a flexion of all the toes occurs. In pyramidal lesions this reflex is abolished and in its place is usually found, in response to similar stimulation of the plantar surface, an *extension* of the great toe. Often with this will be noticed simultaneously a "fanning" or separation of the other toes. The latter, however, is often absent. The essential feature of the phenomenon is a rather deliberate slow extension of the great toe. Except in infancy this phenomenon is always indicative of a pyramidal lesion, although not necessarily a destructive lesion, since mere interference with pyramidal conduction, by pressure or otherwise, may bring forth any of the abnormal reflexes.

It is generally stated that the Babinski phenomenon is normal for the entire period of infancy, on account of lack of development or deficient myelinization of the pyramidal fasciculi. I cannot quite agree with these statements. The more careful the technique the more it will be found that in the average well developed infant plantar flexion is the rule, certainly after the sixth month. There are many factors that lead to error in the attempt to elicit this reflex in the young child. The most disturbing factor is the quick reflex act of defense from the plantar irritation, in which the entire leg is withdrawn by flexion of thigh and leg and dorsal extension of the foot. With the latter the toes also are often simultaneously extended, the extensor proprius pollicis responding more energetically than the extensors of the other toes, with resulting overextension of the great toe. This phenomenon is likely to lead to erroneous assumption of the presence of the Babinski reflex. Another disturbing factor lies in the deceptive appearance produced by a voluntary extension of the great toe. In infants especially, the toes are often in more or less constant movement when the foot is bared. Before the Babinski is assumed to be present repeated tests should be made and the average result of these tests should determine the conclusion. Even then we may be left in doubt. Fortunately in questions of unilateral lesions we have the unaffected side to compare with the supposedly affected side, and in such cases careful comparison of the average results obtained on each side will often reveal the truth.

The extensor response to plantar stimulation is purely a spinal reflex. It is to be considered as a latent reflex, the potentiality of which is always present, but normally held in abeyance by the inhibition of higher centers. This is perhaps true of other abnormal reflexes. In fact the study of the behavior of both plantar reflexes in the presence of lesions of the cerebral motor cortex, or its projection tracts, is one of the strongest arguments in favor of the extraspinal origin of the normal superficial reflexes.

Oppenheim Reflex.—This is merely another method of producing toe extension response in the same conditions in which plantar irritation produces this result. It is performed by drawing some blunt object, such as a pleximeter hammer, down along the inner surface of the tibia with rather firm pressure. It is very unsatisfactory even in adults and much more so in children. When absent no deduction can be drawn from that absence. When present other and more trust-worthy signs of pyramidal lesion are invariably demonstrable. It is mentioned here because so frequently referred to in medical literature.

Gordon Reflex.—Occasionally an extension of the toes may be caused by deep firm gripping of the calf muscles. Attention was first called to this phenomenon by Gordon. I have found it much more dependable than the Oppenheim reflex. Its significance is the same as the Babinski reflex.

Clonus.—Ankle, patellar and wrist clonus. Frequently present in pyramidal lesions is the phenomenon induced by suddenly putting a tendon into a state of tension and holding it there, when successive alternate contractions and relaxations of its muscle occur. The phenomenon is chiefly seen and is most easily demonstrable in the foot, which is grasped by one hand of the examiner while the leg is supported, preferably in a position of slight flexion at the knee, by the other hand. If then the foot is passively dorsally extended by a quick movement, and so held, there ensues a rhythmical rather rapid to and fro movement of the foot, which continues almost indefinitely unless inhibited by some voluntary movement on the patient's part. This is the ankle clonus. It is common but nevertheless frequently absent. Only its presence is significant. Its absence has no negative value.

Under similar conditions a patella clonus may sometimes be elicited. This is brought out by pulling down on the patella with a finger, and, while the tendon is thus held tense, a sharp blow on the finger will be followed by clonic movements of the patella. This is much less common than the ankle clonus.

Occasionally similar clonic movements of a hand may be produced by placing the wrist tendons suddenly under tension.

Strumpell Reflex.—If with the patient in a recumbent position and the lower extremities extended, he is asked to flex one thigh against the resistance of the examiner, a dorsal extension of the foot will occur in certain cases of heightened reflex activity.

Front-tap Contraction.—With the lower extremities in extended position, the foot is passively brought into a position of dorsal extension and the upper outer part of the leg is sharply tapped. If plantar flexion of the foot occurs front-tap contraction is present. There are many other abnormal reflexes but these are the ones commonly employed, and the only ones clinically essential. One or more of these reflexes will be found in every pyramidal lesion.

Complex Reflex Mechanisms and their Disturbance by Disease.—Within the spinal cord lie the sensori-motor pathways and cellular relay stations of a number of special reflex mechanisms whose motor centers are located in the medulla, pons, cerebellum and basal ganglia of the brain. These are therefore spino-cerebral or spino-cerebellar reflexes. Their activity depends upon extra-spinal motor centers.

How intimate is the association between the spinal centers and these intracranial centers is evident when we consider the great system presiding over body equilibrium, muscular coördination, muscle stability and muscle tonus. In this complex mechanism the immediate contraction of the body muscles is unquestionably due to impulses originating within the ganglia cells of the anterior cornua of the cord, but these impulses are excited by and constantly under the control of motor centers in the basal nuclei of the cerebrum and in the cells of the cerebellar cortex, and connected with one another and with the periphery by afferent and efferent pathways throughout the extent of the nervous system.

To keep within the scope of this chapter it is possible only to mention incidentally these spino-cerebral reflex mechanisms, as far as their disturbance may be conspicuously evident in the symptomatology of spinal disease.

Their detailed description belongs elsewhere. There are, however, a number of special reflex automatic functions that may be considered as distinctly of spinal origin, although they also are most intimately associated with the intracranial functions above referred to.

SPECIAL SPINAL CENTERS CONNECTED WITH ORGANIC FUNCTION

Situated in the spinal gray matter at different levels of the cord are a number of nuclei whose duty it is to preside over various automatic functions controlling organic activity. Those which concern us chiefly in the diagnosis and location of spinal disease are those whose disturbance of function by such disease is manifested by conspicuous clinical signs.

Their nature and location is indicated in the accompanying table.

Table 3

SPECIAL SPINAL CENTERS

The phrenic nucleus:	Movements of the diaphragm Inspiratory respiration	C. 3–5
The cilio-spinal nucleus:	Pupillary dilatation	C. 8–D. 1
The thoracic respiratory nuclei:	Thoracic respiratory movements	
Inspiration:	Scaleni muscles	C. 2–8
Inspiration:	Levatores costarum	C. 8–D. 11
Inspiration:	Intercostal muscles	D. 2–11
Expiration:	Intercostal muscles	
Expiration:	Triangularis sterni	D. 3–4
The accessory respiratory nuclei:	Forced inspiration	
	Sternomastoid	C. 2–3
	Trapezius	C. 2–4
	Rhomboids	C. 4–5
	Pectorals	C. 5–8
	Serrati	C. 5–D. 4
	Forced expiration	
	Abdominal muscles	D. 5–L. 1
The genital center		S. 2–3.
The center for the bladder		S. 3–4
The center for the rectum and anus		S. 3–4

Phrenic Nucleus.—The highest of these centers, in the cord proper, is the nucleus of the phrenic nerve, controlling the automatic movement of the diaphragm. The axones of these nuclear cells leave the cord chiefly via the fourth cervical roots, a few filaments coming from the third and fifth cervical roots.

This nucleus is in intimate association with the medullary respiratory centers, with the nuclei of the intercostal muscles in the cervicodorsal cord, with the nuclei of the accessory muscles of respiration, and with the sympathetic system through the lower cervical ganglia and the phrenic plexus. Animal experiments have also shown an apparent connection with the cerebral cortex in the region of the posterior part of the second frontal convolution.

The afferent or sensory portions of the reflex arcs, of which these nuclei constitute the motor division, are too numerous and intricate to be described in detail in a clinical treatise. It is sufficient to point out that the rhythmical automatic movement of the diaphragm must be coördinated with the rest of the respiratory act, in every detail, that while this is going on the mechanism is subjected to the influence of innumerable incoming sensory impulses from all parts of the nervous system, but especially from the nerves of the digestive and respiratory tracts. These impulses may be, and frequently are, so insistent as to inhibit the normal rhythmical outflow of motor impulses from the nucleus, with resultant interference with the normal action of the diaphragm. Such is the act of coughing, due to sensory impulses coming from the naso-pharynx, bronchi, or even from the ear, also the act of sneezing, from irritation of the terminal filaments of the fifth cranial nerve in the nose. In this connection it may be here mentioned that the sensory nuclei of the fifth cranial nerve extend from the pons down as far as the second cervical segment of the cord. These acts are complex reflexes, in which the diaphragm only performs its part.

Hiccough is a phenomenon in which the normal rhythmical contraction of the diaphragm is replaced by spasmodic action. The sensory impulse responsible for it usually arises in the gastro-intestinal tract. The phenomenon may occur, however, from direct irritation of the nuclei or roots of the phrenic nerve and under these circumstances may prove an intractable and serious condition.

Destruction of these nuclei results in paralysis of the diaphragm and leaves the intercostal muscles and the accessory muscles of respiration in sole charge of the respiratory act. If the nuclear cells governing these muscles are in their turn destroyed, the respiratory act ceases and death results.

This may occur in ascending or descending anterior poliomyelitis, in hemorrhage into the gray matter of the cord, or in transverse lesions of the cord, inflammatory or traumatic, in the cervical region. It frequently happens that a crush of the lower cervical cord, from vertebral fracture or dislocation, entirely abolishes intercostal respiration. Under these circumstances the victim lives on for a time, breathing with unimpaired action of the diaphragm. In a few hours, however, a slowly ascending softening, or an extension of the hemorrhage, invariably incidental to such trauma, reaches the phrenic nuclei and sudden death occurs. This contingency must always be kept in mind in all such lesions of the cervical cord.

Cilio-spinal Center.—Another special center is that concerned in the dilatation of the pupils. The reflex mechanism governing pupillary activity is one of the best examples of a complex cerebrospinal and spino-cerebral reflex. In fact, a large part of both the motor and sensory portions of this reflex system lies within the cranial cavity. Nevertheless, the impulses that excite the dilator fibers of the iris originate in a group of cells in the lower part of the cervical enlargement of the cord—the cilio-spinal center—in the eighth cervical and first dorsal segment. Dilatation of the pupils may be excited in many ways. As we have seen, it may exist as a cutaneous reflex in response to irritation of the skin of the cervical region; it may be caused by irritation of the lower cervical and upper dorsal anterior spinal roots, as in meningitis, and it occurs in sympathetic disturbance, but its most important activity depends upon stimulation of the retina. Here the sensory impulses that excite the reflex originate in response to diminishing intensity of light. The impulse passes backward through the optic nerves, optic commissure and lateral filaments of the optic tracts to the superior quadrigeminal bodies, where the impulse is transmitted to cells of their gray matter. From these cells new filaments carry the impulse downward beneath the aqueduct of Sylvius—where the filaments partly decussate—the dorsal tegmental decussation of the anterior tecto-spinal fasciculus—to the cilio-spinal nucleus. On their way down these fasciculi give off filaments to the nuclei of the third, fourth and sixth cranial nerves—the oculomotor nerves. This connection with the oculomotor nuclei is probably concerned with the alterations that occur in the pupils during the act of accommodation for distance.

The pathway to the cilio-spinal center constitutes the middle portion of the reflex arc of the pupillo-dilator tract. The motor portion begins in the cells of the cilio-spinal nucleus and passes out through the anterior roots of the first dorsal segment and joins the cervical cord of the sympathetic—via the rami communicantes, which connect the anterior spinal roots with the sympathetic system—thence to the cavernous plexus, and through the short ciliary nerves to the dilator fibers of the iris. While passing through the cavernous plexus these motor filaments communicate with the nuclei of the oculomotor nerves—probably part of the pupillary mechanism connected with accommodation.

A break in any portion of this reflex arc destroys pupillary dilatation to light. If the break occurs in the portion of this arc that lies between the cilio-spinal center and the dilator iridis absolute paralysis of the dilator iridis occurs and dilatation of the pupils is completely abolished, even to accommodation, otherwise accommodative dilatation is not necessarily lost. When the dilator fibers of the iris are completely paralyzed the sphincter iridis—supplied by the third cranial nerve—is no longer opposed and contracts to its full extent and the so-called pin-point pupil results. Loss of the pupillary response to light with maintenance of response for distance is the so-called Argyll-Robertson pupil. It is seen in para-syphilitic and syphilitic diseases of the nervous system, but chiefly in locomotor ataxia; complete immobility of the pupils being more often seen in paresis and cerebral syphilis. In both of the latter conditions unequal involvement of the pupils is frequently seen, one pupil being affected earlier or more completely than the other, and inequality in the diameters of the pupillary openings is also common, whether they are completely rigid or not. Destruction of the cilio-spinal nucleus, as in poliomyelitis, myelitis, hemorrhage or trauma, or destruction of the first dorsal anterior root close to the vertebra, before these cilio-spinal axones have left it, as in severe lesions of the brachial plexus, results in a contracted pupil on the side of the lesion. This is a not uncommon occurrence in the lower arm type of brachial birth palsy.

Several times cases have been referred to me with a question of the existence of a cerebral lesion on account of this implication of the pupil, a natural mistake if this spinal location of part of the pupillary mechanism is not kept in mind.

When its intricate and intimate connections with other parts of the nervous system are considered, and when the frequency with which it is disturbed by some of the most common of the diseases of the nervous system is realized, it will be seen that testing of pupillary activity is one of the first and most important steps in neurologic examination.

The centers for the thoracic respiratory muscles require no special consideration for their function and location is identical with the nuclei of the several muscles named. They are in constant association with the phrenic nuclei and with many other reflex systems, particularly through the sympathetic mechanism. They are also directly under the control of higher cerebral centers.

The **bladder** and **rectal centers** are situated in the conus. A detailed description of the course of the complex reflex mechanisms responsible for these functions is hardly feasible or necessary. A few facts should be kept in mind, however, in order to understand their disturbance by spinal disease.

They are essentially spinal reflexes* but under the control of higher conscious and subconscious centers. Thus normally, with or without the intervention of the consciousness, these reflexes are inhibited.

As an example we may consider the bladder function. Without higher inhibition the automatic action of this reflex would be as follows. When the bladder became full the pressure against the sphincter and irritation of the neck of the bladder would cause a sensory centripetal impulse to pass through the sensory portion of the reflex mechanism to the spinal center, the sphincter would be relaxed and the muscles of the bladder would contract and the urine would be expelled. As soon as this has occurred and as soon as the bladder is emptied the muscles of the bladder walls relax, the sphincter regains its tonicity and holds back the slowly accumulating urine until distention of the bladder again occurs. This would be the course of the spinal automatic mechanism, acting without interference from other centers. Normally intense pressure of urine under tension within the bladder, and intense irritation of the neck of the bladder, is not able to overcome the strict orders from the higher control to the sphincter not to yield, until time and place are convenient. At least this is what should normally take place when that higher inhibition is once firmly established. In infancy this inhibition has not developed and the emptying of the bladder is a purely automatic act, and throughout early childhood the higher control is easily overthrown and so-called incontinence occurs. This is not incontinence in the true meaning of the term; the bladder-sphincter is perfectly competent and only yields to a perfectly normal reflex impulse. An exactly similar situation exists in lesions of the upper motor segment, when inhibitory control by the higher centers is cut off by the lesion and the bladder mechanism is left free to functionate in a purely automatic way. The conditions then are the same as when inhibitory control is not yet developed, as in infancy.

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* There is no doubt that these reflex mechanisms are in part controlled by lower centers in the sympathetic system, through the hypogastric and hemorrhoidal plexuses, but clinical experience indicates that these lower ganglionic centers are certainly subordinate to spinal nuclei, and not, as Müller assumes, the only centers for these functions.

Müller, Klin. und experiment. Studien ueber die Innervation der Blase, etc. Deutsche Zeitschr. fur Nervenheilk., xxi (1909) S. 86.

Not only is this true in upper segment lesions, but there is often a considerable degree of increased tonicity in the sphincter directly as a result of these lesions so that the sphincter does not readily yield to the reflex impulse of an overfull bladder and *retention* of urine may result. Under such circumstances a few drops at a time may be forced through the rigid sphincter, and a more or less constant dribbling may result. This is sometimes misinterpreted as incontinence and the dangerously distended bladder is overlooked. Such a contingency should always be borne in mind in all instances of spastic paralysis, especially in those of acute onset, and during the stage of coma.

Quite a different condition occurs when any part of the reflex arc for this bladder mechanism is destroyed. Then true incontinence results. The reflex is lost exactly as other reflexes are lost when their arcs are broken.

True incontinence occurs in lesions of the conus and of the cauda equina.

Involuntary emptying of the bladder sometimes occurs in childhood from over-stimulation of the reflex mechanism by a highly acid concentrated urine, or occasionally indirectly by reflex irritation in neighboring parts, as in rectal irritation from oxyuris vermicularis, or irritation from phimosis in either sex, or from vulval or vaginal irritation in the girl. In subconscious states, as in dreams, the reflex may be set in action and micturition occur, and in the unconscious stage of epileptic seizures involuntary emptying of the bladder is common. The possibility of this cause of enuresis should always be thought of in instances of unexplained enuresis, especially the nocturnal form. Many instances of enuresis, however, appear to be due to a lack of tonicity in the vesical sphincter, or to an abnormal excitability of the spinal reflex. It should not be forgotten also that the development of the cerebral inhibitory control depends upon the proper development of the brain and that all conditions preventing or delaying that development are likely to be accompanied by defective control over the lower reflex centers. Enuresis therefore is very frequently associated with mental defect. This should always be thought of whenever incontinence of the anal sphincter is associated with that of the vesical sphincter. The rectal center is also controlled by higher inhibitory centers but the conditions under which the spinal mechanism works are quite different than those that influence the urinary function. The anal sphincter is not subjected to the compulsion of pressure as is the vesical sphincter, and the entire reflex mechanism is much more sluggish and comparatively unresponsive to irritative influence. Involuntary evacuation of the rectum practically never occurs as the result of mere distention. As long as the tonicity of the anal sphincter is intact constipation results. If under these circumstances involuntary defecation occurs it is because the higher inhibitory centers are as yet undeveloped, as in infancy or in the feeble-minded, or have been cut off from the lower spinal centers, as in the case of pyramidal disease. If the reflex arc for the rectal and anal mechanism is broken incontinence of feces may not necessarily result, the rectum may simply fill without any effort being made to expel its contents. It is chiefly when the sphincter ani or its nervous mechanism alone is weakened, while the power of the rectal muscles is intact, that true fecal incontinence occurs. This is not a common occurrence. The incontinence usually referred to is not true incontinence, but merely a spontaneous evacuation of the rectum by normal reflex activity independent of higher inhibitory control. It is what is present in states of coma, in pyramidal lesions irrespective of loss of consciousness, or in lesions of the

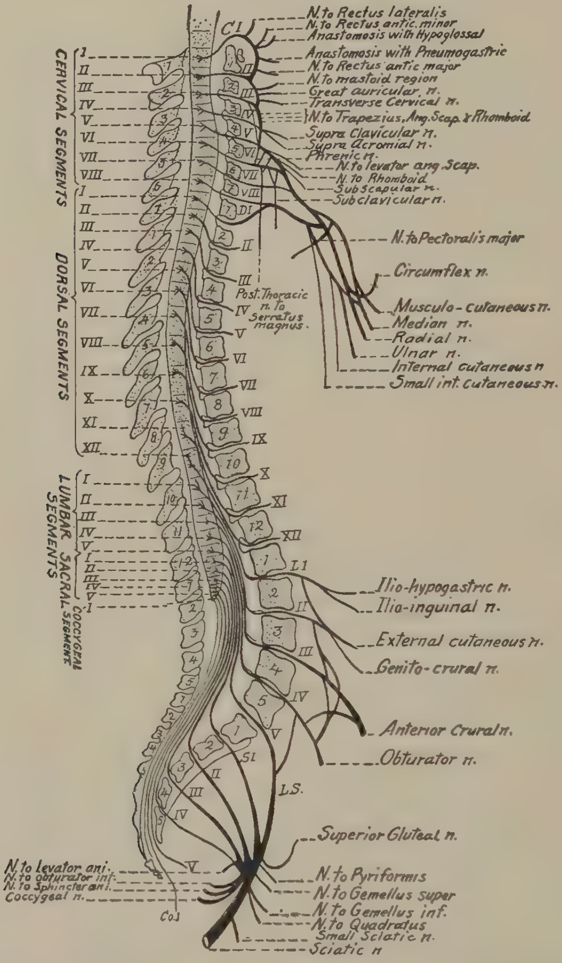

FIG. 70.—The relation of the segments of the spinal cord and their nerve roots to the bodies and spines of the vertebræ (Dejerine et Thomas, Mal. de la Moelle Epiniere, Paris, 1902). (From Starr, Organic Nervous Diseases, 1903.)

cord at any point above the conus, provided the lesion is not one in which there is complete transverse destruction. Genital disturbances as the result of spinal lesions do not particularly concern us during the period of childhood proper, and at the period of puberty such disturbances conform to those that are characteristic of similar lesions in adult life. Even in childhood, however, partial transverse lesions may be accompanied by priapism, as in adults. In the localization of a segment lesion of the cord with reference to the external topography of the spinal column, the fact, referred to in the beginning of this chapter, that the spinal roots corresponding to the various cord-segments do not immediately leave the vertebral canal and do not therefore correspond to the vertical bodies, must be taken into consideration. Moreover our external guide to the various vertebræ are of necessity the tips of their spinous processes which vary greatly in length in different regions of the spine and in the cervical and dorsal regions do not correspond to the level of the vertebral bodies to which they belong but to vertebræ below. Cord lesions therefore are always actually situated considerably above the vertebral level indicated by the spinous processes.

The accompanying illustration (Fig. 70) from Dejerine and Thomas will indicate the relation of the various spinal segments to the vertebral spinous processes and vertebral bodies, as well as the relation of these segments to the points of exit of the spinal nerves. It will be noted that the five sacral segments are crowded in a space scarcely larger than the height of a vertebral body. The long course of the lumbosacral roots is also to be noted.

Lesions of the conus, comprising the three lower sacral segments, are characterized by paralysis of the vesical and anal sphincters, and the bladder and rectal muscles, and loss of sensation in the skin about the anus and adjacent portion of the buttocks, and posterior part of the scrotum or labia. In adults impotence also results.

Destruction of this terminal portion of the cord is usually the result of hemorrhage of traumatic origin.

Lesions of the cauda produce paralysis of the lower neurone type, since it is entirely composed of spinal nerve-roots. Owing to the way in which they are massed together a relatively small lesion is capable of causing extensive symptoms, especially paralysis. This is in rather sharp contrast to the limited extent of the symptoms in a lesion of the conus.

The cauda is formed by the lumbar and sacral roots. The higher roots leave the vertebral canal first and consequently occupy the outer part of the cauda. Their course in the cauda is naturally shorter and they usually suffer less in partial caudal lesions than the lower lumbar and the sacral roots that are obliged to pursue a longer course.

The character of the symptoms are the same as those that occur in root lesions elsewhere, but they are usually bilateral, although practically never quite symmetrical, as is the rule in bilateral lesions of the cord.

The cauda may be injured as the result of vertebral trauma, directly or by the attendant hemorrhage, or it may become involved by disease of the neighboring bone, as in tuberculosis or malignant disease, or it may be implicated in tumors originating in the meninges, or possibly by new-growths within the cauda itself. Penetrating wounds of the vertebræ may injure this as well as any other part of the cord.

DESTRUCTION OF ONE-HALF OF THE CORD: BROWN-SEQUARD PARALYSIS

Destruction of one-half of the cord at a given level produces the characteristic clinical picture known by the above title. When present in its complete form the symptoms are as follows:

On the side of the lesion: Flaccid paralysis, corresponding to the level of the lesion; spastic paralysis of all muscles below; a band of anesthesia for all forms of sensation, corresponding to the sensory roots destroyed; above and below this band a zone of hyperesthesia, corresponding to the root-fibers that touch upon and are irritated by the lesion but are not destroyed by it; a diminution of muscle-sense and of the sense of position of the limb below the lesion.

On the side opposite the lesion: Complete anesthesia for touch temperature and pain below the level of the lesion up to about an inch below the level of the anesthetic band of the other side. Just above the level of this anesthesia is a single band of hyperesthesia corresponding to the irritated fibers of the posterior root that cross to the other side of the cord and just graze the top of the lesion. The tendon reflexes may be somewhat increased but there is no paralysis on this side.

In addition to the above well defined symptom-complex there is sometimes found on the side of and below the lesion a certain degree of hyperesthesia to touch, temperature and pain. It is not constant.

The absolutely typical picture above described is perhaps less common than a modification of it, but at all events it is a condition easily recognized. The essential features are some degree of the spastic type of paralysis on the side of the lesion and the sensory disturbance on the opposite side, both below the lesion.

The condition is usually secondary to stab wounds or bullet wounds of one-half of the cord. I have see a somewhat similar syndrome as the result of hemorrhage into the cord but without the hyperesthetic areas, and an instance in an adult secondary to fracture of the fourth cervical vertebra, with implication of the phrenic nucleus. The hyperesthetic areas are not often a conspicuous feature of the condition.

From the foregoing description of the functions of the various portions of the spinal cord it is evident that the vertical or segmental localization of a spinal lesion depends upon the presence in the symptom-complex of certain phenomena, the existence of which can only be explained by interference with the functions of the cord at a particular level. These phenomena consist of motor and sensory disturbances and occasionally of disturbance of special organic function. The localizing motor disturbances are of the lower motor neurone type, namely flaccid paralysis with loss of the reflexes whose sensori-motor arcs correspond to the level of the lesion.

The successful employment of these methods of localization depends largely upon actual clinical experience, and especially on the skill with which the examination is conducted and deductions drawn from its result.

The citation of a few cases will perhaps supplement the foregoing outline of spinal localization. While I have selected almost exclusively instances that have come under my observation in children, the same principles apply to cases at any time of life. For the sake of clearness the cases selected have been chosen from among those in which the localization is clearly defined. It should be borne in mind that the particular type of clinical picture seen in early cases may be quite altered by the subsequent course of the case and the following case is a typical illustration of this. The early indications were those of a complete transverse destruction, but the subsequent picture was that of injury chiefly of the pyramidal tracts.

Child 2½ years old: She was struck across the back by the fender of a trolley car 10 days previously. There was complete flaccid paraplegia with total loss of sensation below the level of the umbilicus, loss of the lower abdominal reflexes and of all the reflexes in the lower extremities, urinary retention, involuntary evacuation of the bowels. Radiograph negative. Cystitis was present, also hematuria during the first week.

The diagnosis was of a hemorrhage into the cord at the ninth and tenth dorsal segment.

Very slight movement in the toes suggested that the destruction was not complete and the subsequent history proved the assumption to be correct.

Very slow subsidence of the flaccidity occurred, and at the end of several months exaggeration of the deep reflexes in the legs with double ankle clonus and bilateral Babinski were present. The child was however entirely helpless below the waist. Unfortunately such improvement as occurred in this case was of no functional value.

The following case is an example of complete destruction of the cord at and below the level of the first lumbar segment.

Child 12 years old: While swinging, with other children, in a hammock attached to a chimney, the chimney fell crushing her under the debris. There was total flaccid paraplegia with abolition of all reflexes in the lower extremities, complete anesthesia below the level of Poupart's ligament, and involuntary evacuation of urine and feces. She showed no improvement when last seen.

The following is an interesting example of bilateral pressure on the cord, with symptoms limited to the pyramidal tracts. As has already been mentioned, it is impossible to accurately localize such a lesion unless there is accompanying sensory disturbance or bony deformity.

Child 11 years old: Fell, a week ago, from a veranda to the ground, 20 ft. below. When picked up, two minutes later, he was conscious. There was clonic spasm in both legs which lasted two days, leaving the legs paralyzed. There was no incontinence. He complained of pain in the upper abdomen and in the mid-dorsal region. He presented an intense spastic paraplegia, with great exaggeration of the tendon reflexes, double ankle clonus, double Babinski and Oppenheim reflexes. There was intense hyperesthesia in the epigastrium, and a band of hyperesthesia, 3 in. wide around the body corresponding to the sixth to the ninth dorsal segments. Above and below this band all forms of sensation were normal. There was intense tenderness on pressure over and pain on movement of the lower dorsal spine. Diagnosis: Dislocation of the tenth dorsal vertebra.

Under ether this was reduced, and the boy made a complete recovery, showing a month later only somewhat exaggerated knee-jerks.

Child four years old: Shot by a playmate. Exit of bullet 1 in. to left of the spinous process of the eleventh dorsal vertebra. Displacement of the spinous process of the twelfth dorsal vertebra.

Spastic paraplegia with diminished sensation from level of umbilicus downward. This case was seen some time after the infliction of the injury and the condition was apparently stationary and permanent. In this case the cord must have been injured at about the level of the tenth dorsal segment, much higher than the exit of the bullet would indicate.

TABES DORSALIS*

(Posterior sclerosis, locomotor ataxia)

Incidence.—This disease, one of the most common of organic nervous diseases of adult life, is one of the rarest in childhood. In 25 years of practice I have not seen an instance within the period of childhood, although for many years connected with two large neurologic clinics, one of which is exclusively devoted to the nervous diseases of children.

It is hardly necessary to say that I have been keenly on the watch for incipient symptoms of the disease and that I feel confident that no such instance has been overlooked.

In over 7000 consecutive cases of nervous diseases in children, at our clinic at the Children's Hospital, not a single instance of tabes has appeared, although a large number of cases of ordinary cerebrospinal syphilis have been under observation there.

Among all the tabetics seen at our clinic at the City Hospital not a single instance of this disease in childhood has occurred. When all doubtful cases are eliminated the entire medical literature furnishes but 60 instances of the occurrence of tabes in the period of childhood or previous to the fifteenth year. Cases of tabes from inherited syphilis appearing after puberty are considerably more numerous, as a number have been reported with the onset subsequent to the fifteenth year.

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* The description of this disease applies only to its manifestations in children, and is based entirely on a study of all available clinical material within the period of childhood. The symptom-complex, therefore, represents such phenomena as have actually occurred in cases that have been reported in children.

I have no hesitation therefore in stating that tabes is rarer than any other disease of the nervous system in childhood. One reason for its rarity might seem to be in the long period usually elapsing between the initial infection and the beginning of the clinical signs of tabes, but among many hundreds of adult tabetics I have seen but one instance that seemed of hereditary origin. Brun's suggestion that its rarity may be due to the early mortality of hereditary syphilitic children does not hold good, for in that case cerebral syphilis itself would be rare in childhood, which is not the fact. Moreover the number of hereditary syphilitic children surviving to puberty and beyond is sufficiently large to furnish a considerable number of cases of tabes provided that the same liability to the disease existed in hereditary syphilis that exists in acquired syphilis.

Paralytic dementia (general paresis) is much less rare, although very uncommon. I have seen but three instances in children, of which two came to autopsy. Certain cases reported in the literature as instances of infantile tabes are so classified on insufficient evidence, for some are clearly instances of cerebello-spinal ataxia ("hereditary ataxia") and others of ordinary cerebrospinal syphilis. The list of such instances is a long one and cannot be critically considered here.

Etiology.—Like paralytic dementia there is but one etiologic factor in this affection and that is syphilis. It is frequently referred to as a parasyphilitic or metasyphilitic disease. In the light of our present knowledge, however, these terms have but little meaning. It is now known that the treponema pallidum is present in both paralytic dementia and tabes. Both of these affections are in reality the manifestations of actual syphilitic disease. It is probable that the virus in both of these instances, although perhaps more especially so in the case of tabes, is in a state of attenuated activity or vitality. Of 57 cases that may be considered unquestionable instances of tabes commencing within the period of childhood, 45 were due to hereditary syphilis, four were definitely ascertained to be due to acquired syphilis, and in eight cases the time of infection was not definitely determined. Of special interest among the hereditary instances are those in which one or both of the parents suffered from tabes or paresis. I have called attention to a similar fact in connection with a study of paresis in childhood.*

Thus among 57 children afflicted with tabes there were 25 parents who were victims of tabes or paresis.

Even more remarkable are those instances in which more than one child in a family becomes tabetic, and Nonne† cites an instance in which two daughters of a tabetic mother became tabetic, and Stiefler has recently reported an instance of two tabetic children in a family of seven all suffering from acquired syphilis, the father being also tabetic. These facts, as well as the occasional occurrence of ordinary cerebral syphilis in the parents, points to the probability of familiar predisposition to the disease, or at least an unusual susceptibility of the nervous system in certain families. Considering the rarity of infantile tabes the development of two cases in one family could hardly be otherwise interpreted. Contrary to the rule among adult tabetics, where male victims are greatly in excess, infantile tabes afflicts both sexes about equally.

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* Fairbanks, General paresis in childhood, Journ. Amer. Med. Assoc., li (1908), p. 1946.

† Nonne, Syphilis des Nervensystems 2 Auflage, Berlin (1908).
Stiefler, Ueber fam. u. juven. Tabes Wien. kl. Wochensch., xxii (1909), p. 163.

Symptomatology.—The cases on which this study is based all commenced before the termination of the fifteenth year, and may therefore be considered as strictly instances of the affection actually occurring in childhood.* The number is sufficiently large, and the clinical data sufficiently complete, to justify the presentation of a symptomatology of the disease as it manifests itself at this period of life. Moreover a comparative study of the clinical phenomena in these cases in childhood, with the adult clinical picture in mind, shows a rather striking uniformity in the features of the symptom-complex, in childhood, a uniformity that warrants the conclusion that this disease presents a picture *sui generis* in the early years of life. Nevertheless the clinical picture is perfectly typical. It is we who must change somewhat the perspective acquired by our familiarity with the disease as it manifests itself in later life. This is particularly true of the affection in its incipient stage.

Initial Symptoms.—These, in the order of their frequency of occurrence, and their percentage of the number of cases in which the initia symptom was determined with reasonable certainty, are as follows:

	Per Cent.
Failing vision	30
Disturbance of bladder function	30
Sensory disturbances	20
Pupillary changes	6
Mental deterioration	6
Visceral crises	4
Diplopia	2

It is probable that failing vision is even more often the first sign of the disease than these figures indicate, for it is frequently not detected in its early stage in the child.

It is due to optic atrophy. As is well known a considerable degree of alteration in the nerve may exist without commensurate involvement of subjective visual acuity. This was evident also in some of the cases in childhood, in which pallor of the disc was noticed before visual disturbance appeared. This occurred in four out of 48 cases in which the fundus was examined. If these cases were included the above percentage would be higher. This incidence of optic atrophy as an initial symptom of tabes in childhood is five times greater than the incidence of optic atrophy in adult life throughout the entire course of the disease. Next in frequency as an initial symptom is some form of bladder disturbance, usually incontinence, either diurnal or nocturnal or both, but occasionally difficulty in emptying the bladder occurs.

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* There are approximately 60 such cases on record in which there is no reason to question the diagnosis. I was unable, however, to obtain the clinical details in the remaining cases to an extent that warranted their utilization statistically.

Incontinence may take the form of complete sphincteric relaxation, with complete emptying of the bladder, or it may occur in the form of "dribbling" when the bladder is full and the sphincter is unable to hold perfectly against the pressure. Occasionally the complaint is the necessity of haste in emptying the bladder lest spontaneous evacuation occurs.

These bladder, or sphincteric disturbances are also common early phenomena in adults.

Initial sensory phenomena fall far below visual and sphincteric disturbances in frequency, yet they are fairly common. It is chiefly when we consider that they head the list of incipient symptoms of adult tabes, that their relative infrequency in childhood becomes conspicuous. They take the form in the child, as in the adult, of stabbing or boring pains, of more or less paroxysmal character, usually in the lower extremities but sometimes in the arms. Paresthesia in the onset of tabes in the child is very rare, only a single case being on record.

Of the remaining initial symptoms of tabes the one of chief importance is change in the mental state. When such change occurs as an initial phenomenon it indicates that the case has commenced as a paresis, whatever the subsequent course may be. The early diagnosis of such instances may be obscured by the occurrence of convulsive attacks or possibly by a hemiplegic attack. These instances must be carefully differentiated from ordinary cerebral syphilis.

Of much interest, although great rarity, are those cases that first make known their presence by a visceral crisis. As might be imagined the sudden onset of violent abdominal symptoms in a child of six, or a child of 11, would hardly suggest a tabetic origin, yet this was the case in two instances.

Probably the earliest sign of tabes appears in the alteration in size and in response of the pupils to light, but this is not usually detected until the case comes under observation for other and later symptoms, so that any attempt to determine its frequency as an initial phenomenon is impossible.

Diplopia, a frequent early occurrence in the adult, is extremely rare in childhood.

Before passing to a consideration of the general symptomatology of the disease in its subsequent stages, one symptom, comparatively common in the early stage of the disease, should be mentioned. This is headache. It is not justifiable to include it as an incipient phenomenon for it is a symptom of so many conditions that it may have no relationship whatever to the onset of the tabetic process.

We may now proceed to a consideration of the phenomena of the subsequent stages of the disease, and it should be borne in mind that all of the common initial symptoms are also conspicuous features of the clinical picture presented by the fully developed disease. Thus, whereas we have seen that atrophy of the optic nerve appears as an initial symptom in nearly one-third of the cases, we find that it is a conspicuous feature of the general symptom-complex in two-thirds of the cases. Here we see an even more remarkable variation from the clinical history of adult tabes. This has evidently no special connection with hereditary syphilis for it was not only present in three out of the four instances of acquired syphilis but it was the initial symptom in all three cases. It is a characteristic of tabes developing in childhood.

Sensory Symptoms.—The sensory disturbances are due to involvement of the posterior nerve roots. They comprise phenomena due both to irritation and to destruction of these roots. Irritative symptoms comprise spontaneous pain, paresthesic and "girdle" sensations. Some form of irritative root symptoms occurs in over half the cases. Of these symptoms pain is the most prominent. It is usually felt in the lower extremities, sometimes also in the upper extremities, almost never in the arms alone. It may rarely appear in the trigeminal, and in one instance was limited to that nerve. It is distinctly paroxysmal, lancinating or boring, and of varying duration, usually momentary, but soon recurring. It may be severe for several days, or weeks, then disappear for considerable periods, or it may occur with greater or lesser severity for a year or more and then cease and not again recur. On the whole the pains do not constitute in childhood the element of extreme suffering so often seen in adult life. Occasionally a dull aching in the legs is a source of discomfort. Paresthesia is much less often complained of in childhood. It was a feature of one-quarter of the cases in which investigation of sensory phenomena was made. It takes the form of complaint of numbness, coldness, prickling and creeping sensations, usually in the feet, and occasionally of the sensation of "walking on pillows" or velvet.

The paresthesic sensations may be constant but are more frequently intermittent. Rarely peculiar attacks are seen. In these a sensation of numbness or tingling commences in toes or fingers and ascends the limb, associated with a feeling of weakness in the extremity affected. The fact that these attacks affect one-half of the body and have been accompanied by headache, vomiting or involvement of speech, would suggest that they are of central origin. In the instances in which they occurred, however, there were no other cerebral signs and the tabetic process had existed for several years. Duration of the attacks from a few minutes to an hour. Both the spontaneous pain and the paresthesic phenomena may be unilateral at first, but sooner or later usually appear in the corresponding extremity of the other side.

When present, the paresthesia appears usually coincidently with or soon after the onset of the root pains.

Girdle sensations are not common in childhood, or perhaps it would be more accurate to say that they are not often mentioned by children. They are a form of paresthesia, and such sensations are not noticed in early life in the same degree that pain attracts the attention of the child.

The sensation is defined as a feeling of constriction or "tightness" about the body.

Destructive sensory phenomena consist of varying degrees of diminution or of loss of the different forms of sensation in any region of the body.

The most common disturbance of sensation is some degree of diminution or a loss of the perception of pain. It seems to be more than twice as common as impairment of tactile sensation. It is quite possible, however, that this is because it is much more easily tested in childhood than the sense of touch. It is more frequently to be demonstrated in the lower extremities than elsewhere, but it is not uncommonly found on the anterior surface of the thorax, at or below the mammary region, less frequently in the arms, occasionally in the face. It is more often diminished perception of pain rather than absolute loss.

Diminished tactile perception, or complete anesthesia, is also more frequently found in the lower extremities, especially in the feet, but not rarely on the anterior thorax. It is much less commonly detected in the arms and on the face. Implication of the temperature sense has been tested so infrequently that no conception of its relative implication can be formed.

Involvement of deep sensation has been found in but two instances in childhood.

Delayed perception of pain has also been twice reported.

Both of these disturbances of sensation are probably more common than these figures indicate. In fact this statement is probably true of all sensory disturbance. In this, as in other diseases, it is extremely difficult to obtain reliable response to sensory tests in the child.

The areas of sensory involvement correspond to the territory supplied by definite spinal roots, except in those rare instances where an actual peripheral neuritis exists, in which case the location corresponds to the cutaneous distribution of the particular nerve involved. When neuritis occurs it is usually located in the branches emanating from the brachial plexus, especially the musculo-spiral or the ulnar nerve.

Ataxia.—Considering the comparative frequency of sensory disturbance the relative infrequency of disturbances of coördination for finer movements is a striking fact. In the cases in which this symptom appears to have been investigated it occurred in only 31 per cent., and in many of these instances it was only slightly pronounced. It appears to be a late phenomenon in practically all instances, and the above percentage indicates only those cases where it appeared before the completion of the fifteenth year. Nevertheless even with the inclusion of later developing ataxia, it is a relatively infrequent symptom of tabes commencing in childhood. It has been thought that the infrequency of ataxia is especially associated with the occurrence of early optic atrophy.

Of 25 cases where *early* optic atrophy was well pronounced 60 per cent. were without ataxia symptoms, while 40 per cent. showed ataxia of some degree, notwithstanding the severe involvement of the optic nerve. Of the cases without optic atrophy, 78 per cent. were also without ataxia, while 22 per cent. showed ataxia with normal vision. These figures are of interest but hardly justify any sweeping generalization. Certainly the study of the disease in childhood does not confirm Benedikt's axiom that ataxic symptoms disappear or fail to develop after the appearance of optic atrophy, nor Gower's opinion that cases with early optic atrophy often fail to develop the late symptoms of tabes. Whatever the reason may be it may be definitely concluded that ataxia is a very inconspicuous feature of the symptom-complex in childhood. Closely connected with the ataxia, and sometimes preceding it for several weeks or months, is a complaint of fatigue on exertion. It is also a common occurrence in ataxic conditions in childhood other than tabes.

Disturbance of Station and Gait.—Much more common than ataxia is disturbance of equilibrium. It occurred in the form of Romberg's phenomenon in 50 per cent. of the cases. It is often slight in degree however. Occasionally the sense of defective balance, in the absence of visual aid, is subjective, when it may not be objectively evident. This is also the case among adults, and is true of any disturbance of equilibrium regardless of its cause, especially in its early stage. Objective disturbance of locomotion appears to be very uncommon, and even a subjective sense of difficulty is not often a source of complaint. When noticed by the child it is described as the interference of one foot with the other. This is due to ataxia in the legs. The disturbances of gait due to Romberg's phenomenon ought to be common in view of the frequency of blindness due to optic atrophy. This is however not the case, and this fact opens up interesting speculation as to the influence of the intensive training of the paths concerned in equilibration, necessitated by the early loss of visual aid, on the non-appearance of this symptom, that is so conspicuous a feature of adult tabes.

The ataxia for finer movements of the extremities and the disturbances of station and gait in tabes are the result of the brain's failing to receive, through the usual channels, proper information of the status of muscles and joints, and the consequent position of the limbs, at any given moment. This information is normally transmitted through the peripheral nerves, posterior spinal roots and posterior columns of the cord, in other words entirely through the structures in which the tabetic process is located. The brain is obliged therefore to rely upon vision for the information usually obtained by way of those paths. The sudden deprivation of visual aid by closing of the eyes therefore results in the disturbances above referred to. For the same reason the ability to determine the weight, form and size of objects is affected. No investigation of these varieties of disturbance of muscle sense has been made in childhood.

The cause of the ataxic disturbances of tabes is a lack of that fine perception of the exact degree of muscular force requisite to perform a given act. Consequently voluntary acts are overdone and a sought-for goal is over-reached.

As we shall see when we come to consider the treatment, it has been found that constant practice will overcome to a great extent the disabilities here referred to.

Cranial Nerve Involvement, Other than the Optic.—*Pupillary Phenomena.*—These form the most constant of the objective signs of the disease. They are probably present at some time during the course of the affection in all cases, and are usually very early signs, but no actual estimate of the percentage can be given for, incredible as it may seem, several cases of tabes reported by oculists have no mention whatever of the condition of the pupils.

The most common disturbance consists of a gradual loss of response to light with more or less maintenance of response to accommodation, the so-called Argyll-Robertson pupil. This occurs in about two-thirds of the cases in which the pupils are implicated. In the remaining third, response to accommodation is also involved, either entirely lost or sluggish. Either condition may be noticed in one eye before it becomes sufficiently marked to be noticeable on the other side, but this is not very often the case, because the condition is usually of considerable standing by the time it is first discovered. The pupils may be fixed in dilatation or in contraction, but dilatation of some degree is by far the more common of the two, as it also is in adult life. A pupil apparently fixed may occasionally respond to light directed into the other eye—consensual response.

Inequality of the pupils is very common. On account of lack of uniformity in methods of examination by various observers its relative frequency in childhood cannot be stated, but it was noted in about half of the cases reported.

Oculomotor Nerves.—Involvement of the external ocular nerves is not common. When it occurs some one or more of the branches of the third nerve are those usually implicated, especially the branches to the levator palpebræ superioris with resultant ptosis, and those to the internal rectus with external strabismus, or at least with sufficient overthrow of muscular balance to cause diplopia. In one instance two years after the onset at nine years, there occurred left ptosis, diplopia and failing vision, and later on double abducens paralysis occurred. This case was under continuous observation for 13 years.* The diplopia is usually transitory, although it may be troublesome for some time.

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* Remak's case. Under Mendel's observation for 10 years. It was unquestionably a case of tabes.

Trigeminal involvement has already been considered, at least as far as its sensory portion is concerned. There is no record of disturbance in its motor portion.

Facial involvement is not yet reported in childhood, and the same is true of the auditory nerve. It is probable that some degree of implication of this nerve would be found if more often searched for. Its implication is certainly not uncommon in the adult. The same may be said of labyrinthine disturbance. Two instances of tabes commencing in childhood are reported as showing facial implication in later years, and in one of these the tenth and twelfth were also affected. This was an instance of multiple bulbar palsies.*

Bladder disturbances, as might be anticipated, are extremely frequent. As we have seen, they constitute a large proportion of the initial symptoms of the disease, and their nature has been in the main already described and requires no repetition here. Instead of incontinence in its various forms, dysuria may occur and considerable difficulty in emptying the bladder may be experienced. There is often an imperative desire to urinate without the ability to immediately satisfy the desire. This may appear early or it may supervene after some years of incontinence, or it may alternate with incontinence, or it may exist with constant dribbling, between times, by day, due to over-filled bladder, which by night, during sleep, is spontaneously able to evacuate its contents.

Incontinence of feces is fortunately extremely rare. It occurred in Remak's case in conjunction with urinary incontinence following three years of dysuria, at the age of 12, in the third year of the disease.

Visceral Crises.—These occurred in 14 per cent. of the series of cases studied.† They comprised gastric and intestinal phenomena ranging from the severest to the lightest forms. The mildest type is the appearance of attacks of apparently causeless vomiting either alone or attended by epigastric pain and more or less general prostration. The severer attacks are ushered in by intense nausea and a sense of epigastric pressure or intense burning sensation, soon followed by incessant vomiting, at first of food, then of bile and mucus or large amounts of watery fluid, lasting from a few hours to several days, then disappearing completely, leaving usually intense thirst. The vomitus may be of the coffee-ground variety. During the attacks the stomach is absolutely intolerant of fluids or solids. In the intervals between the attacks the victim is free from gastric symptoms. Even immediately following an attack food and drink may be tolerated. Great prostration, however, persists for some hours. In the beginning of short duration with long intervals between, they tend to occur at more frequent intervals and to lengthen in duration and increase in severity. They may, however, disappear for long periods. The intestinal crises are characterized by attacks of diarrhea with or without pain. In adults the writer has seen a good many cases in which operations for appendicitis or gallstones had been performed, needless to say without a preliminary neurologic examination. The relative frequency of gastric crises in childhood makes it probable that pharyngeal, laryngeal, vesical and other crises will in time be reported unless the incidence of syphilis itself in early life is greatly reduced.

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* Hudovernig's case.

† This is considerably greater than the average incidence in adult tabetics.

Reflexes.—The deep tendon and bone reflexes are diminished or abolished at all levels where the process of degeneration in the posterior roots has reached a degree of severity sufficient to break the sensory portion of the reflex arc. As the tabetic process in the majority of cases commences in the dorso-lumbar cord, the patellar reflex is affected at a very early period. Next to suffer is the achilles reflex. The condition of these two reflexes therefore furnishes one of the most valuable early indications of the disease.

The tendon reflexes of the upper extremities are far less frequently affected, which is what is to be expected considering the usual location of the disease.

The involvement of these deep reflexes is often not quite symmetrical and may be evident on one side before it appears on the other side.

The superficial reflexes enjoy long immunity, a fact which as yet is unexplained. Late in the disease they also may disappear.* Hypotonia was reported in seven of the cases in childhood, about 12 per cent.

Mental Disturbances.—The unclassifiable mental disturbances, not infrequently seen in adult tabetics, do not seem to occur in childhood. The mental phenomena that do occur in early life are such as are to be explained on the basis of a co-existent paretic process. This process may remain subordinate throughout the course of the disease, or it may at any time rapidly increase so as to eventually overshadow the tabetic phenomena.

Convulsive Attacks.—These are not common. When they occur they also should excite suspicion of a smoldering leptomeningeal process, although this may never assume a conspicuous place in the symptom-complex. Indeed no other symptom may appear.

Hemiplegic attacks are to be regarded also as extremely suggestive of a paretic process combined with the tabes, and secondary to cerebral arterial disease. They are very rare.

Scoliosis, dating from early childhood, has been reported in several cases. What relation it bears to tabes, or whether its presence is only a coincidence, is not apparent.

The remaining symptoms of tabes, some of which are classical, in the adult, are practically unknown in the child. Thus I find but a single instance of trophic disturbance in childhood, a perforating ulcer of the foot in a child of 15, of which the age of onset is not stated, and I find no instance of arthropathy occurring before the end of the fifteenth year. There is no record of muscular atrophy or other evidence of implication of the anterior horns or nerve-roots. It is of course a relatively rare occurrence in adults. Physical deficiency has been present in a considerable number of these tabetic children. This introduces the interesting problem of the effect of the disease on a developing organism. In the adult the deleterious influence of the disease on the nutrition of its victim is a common clinical fact. It is not surprising therefore to find it even more deleterious in its effects on the organism in an active period of development.

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* Attention is again called to the precaution to be observed in the testing of reflex phenomena in children and in the interpretation of the results.

The frequent mental retardation may be explained on similar grounds. Indeed the effect of early syphilis alone on both physical and mental evolution is well known.

With the above consideration of the predominant symptoms of tabes in childhood and in view of the rarity of the disease it is not necessary to attempt to present a clinical picture of the order of appearance of the phenomena. It is essential to keep in mind the initial signs of the disease, as they have appeared in these cases, and to remember that the disease is insidious in its onset and slow in its course, and that after the appearance of its initial symptoms there is no rule as to the time of onset of the other phenomena. They may appear in any order.

In a general way it may be said that the clinical picture presented by the disease is perfectly typical but on the whole one of greater simplicity than that seen in adults, and that its course is milder.

Pathology.—The only instances of the disease in childhood that have come to autopsy are the cases of Westphal and Köster. These were both instances of tabo-paresis, and the pathologic pictures presented by both the brain and cord were typical of the two processes. Very careful and detailed examinations of these two cases are on record. The condition of the cord in each instance does not differ from that seen in the disease in adults. There is no reason to suppose that more extensive pathologic material in childhood would show any material difference from the alterations found in later life.

These changes consist of a degeneration of any or all of the axonic fibers that enter the spinal cord through the posterior nerve roots. From these incoming fibers are formed the columns of Goll and Burdach, the marginal zones of Lissauer, the comma tracts of Schultz, the bundles of fibers traversing the root-zones to the dorsal nucleus of Clarke, and other nuclei of the posterior, central and anterior gray matter of the cord, the latter probably being reflex paths. These fibers are all concerned with centripetal sensory impulses, and all pass through the posterior spinal root ganglia.

Perhaps no other problem in pathology has been more discussed or has given rise to more patient investigation than that of the site of origin of the tabetic process. It is not possible, within the limits permitted in this chapter, to present the merits and demerits of the ganglionic, the meningeal, the neuritic, the vascular and the lymphatic theories of the origin of this disease. Certain of these theories have appealed strongly to many writers, and perhaps none more so than the theory of the ganglionic origin of the process, and there has been an increasing tendency towards its acceptance during the last few years. A critical study, however, of the character and degree of the histopathology of all the available cases in which careful examination by modern methods of staining has been carried out, does not support this theory. There is no question as to the occurrence, in certain instances, of widespread degenerative changes in the spinal ganglia, or in other cases of the presence of such changes in some of the ganglia, but these changes do not possess the intensity, the uniformity, or the invariability requisite for their acceptance as the primary location of the process, to which all other changes may be regarded as secondary. Not only do many instances of tabes of intense degree and widespread extent show no ganglionic alteration, but in such as do present ganglionic alteration the axonic degenerative changes within the ganglia are not what would be expected in primary destruction of a sensory cell. This degeneration is practically limited to the central or spinal branch of the cell and even this degeneration is not always commensurate with the severity of the process in the corresponding posterior roots. Even less frequent and less consistent with the intensity of the process in the posterior roots are the alterations that have sometimes been found in the ganglionic cells, and it is very probable that where these cellular alterations are found they are secondary to degenerative processes in the peripheral axone of the cell—peripheral nerve degeneration—as indicated by the experimental investigations of Lugaro. However, the subsequent investigations of Köster and Kleist seem to indicate that similar cellular changes occur, only after a somewhat longer period of time, when the cell is cut off from its spinal or central connection. The experiments of these three investigators indicate that any changes that occur in the ganglia may well be secondary to the degeneration of the axones outside of the ganglia either peripheral or central.

Gross alterations in the spinal meninges are neither sufficiently intense nor sufficiently extensive to account for the degenerative changes of this disease, nor are such alterations invariably present. Moreover, no similarly distributed degeneration is found in diseases other than tabes in which well-pronounced meningeal infiltration is invariably present. Vascular changes within the meninges will be referred to later. Nageotte is responsible for the root-neuritis theory —névrite radiculaire interstitielle transverse—based on careful histologic examination in 11 cases of tabes. This process, according to Nageotte, consists primarily of a perineuritis, which later extends into the intrafascicular connective tissue and becomes an endoneuritis. The process is limited to that portion of the spinal roots that extends from the point of union of posterior and anterior roots nearly but not quite to the spinal ganglia, a small section intervening in which the process is not conspicuous, especially in cases not of long duration. Early cases show changes only in the middle part of this portion of the spinal roots. There is no question but that these changes do occur, in tabes. They are in fact common although they have not been found by all investigators who have examined this portion of the posterior roots. Equally important is the fact that they are found early in the course of the disease and are then often confined to the middle portion of this particular segment of the spinal roots, gradually extending thence in both directions along the course of the roots. Nageotte was inclined to regard the process, as secondary to a meningitis. It seems to the writer that he erred in emphasizing the neuritic nature of the alterations found, about and within the nerve roots, and in laying too little stress upon the vascular changes in the meninges covering the nerve roots, although he casually mentions such changes. It seems probable that some of the alterations found may indicate a certain degree of inflammation but that this is merely incidental to the inflammatory process going on in and about the small vessels of the pial sheath and their extensions into the intrafascicular connective tissue of the nerve roots. For many reasons impossible to give in detail here this assumption best explains the tabetic process. Under this hypothesis the process is to be regarded as primarily a parenchymatous nerve degeneration of vascular origin, with a replacement proliferation of connective tissue, in which however inflammatory elements may occasionally be seen as the direct result of vascular inflammatory processes. This portion of the spinal roots is invested with a sheath of pia-arachnoid and dura. The pia-arachnoid is thickened and its vessels show well-marked changes, consisting of a thickening of the walls and a diminution in caliber, resulting, here and there, in complete obliteration of the vessel. Shäffer has rightly laid stress upon this fact. In the writer's opinion the infiltration of lymphocytes and plasma cells in the perivascular so-called lymph-spaces in the vessels of the pia and their branches in the connective tissue of the cord, found by Schröder in five cases of tabes, represents merely the vascular inflammatory process referred to. Obersteiner and Redlich have placed the commencement of the tabetic process at that point where the posterior roots enter the cord. Here there is normally found a constriction in the roots and the medullary sheath is thinner than elsewhere. Emphasis is laid by these authors on the theory of a shrinking of the pia through interstitial contraction with consequent injury of the root fibers.

It seems to the writer from an extensive survey of the work thus far accomplished in the histopathology of the disease and a critical weighing of all the facts, that the vascular theory of the tabetic process has the weight of evidence in its favor, that the process is due to vascular changes in the minute vessels of the pia, and that the ischemic atrophy resulting from the blocking of the circulation commences in some instances in the extramedullary portion of the posterior roots, in other instances in the root-zones, or at the point where these roots enter the cord and that the degeneration of other tracts in the cord is to be accounted for on similar grounds, and that the slow extension of the process from the posterior to the lateral region of the cord, and sometimes the involvement of the anterior region is due to the gradually extending obliteration of the minute vessels supplying these sections of the cord. The almost universal beginning of the process in the lower cord, as well as the isolated involvement of certain fibers far distant from the seat of the principal degeneration, cannot be explained satisfactorily on any other premise. For this vascular process a meningitis in the strict sense of the term is not essential, although not infrequently present. Indeed the practically invariable lymphocytosis, in the cerebrospinal fluid in tabes indicates that some degree of meningitis is probably always present.

To resume consideration of the general pathology of the disease it may be said that while tabes presents its characteristic changes in the tracts enumerated at the commencement of this section, certain changes are sometimes found elsewhere in the spinal cord and even in the peripheral nerves. Among these changes one of the most common is more or less degeneration in the fibers of the pyramidal tract. This was, for instance, found in both of the cases in which there was opportunity to examine the cord in tabetic children. These were both instances of tabo-paresis and such degeneration might be anticipated as the result of the cortical degeneration invariably found in paresis. Undoubtedly some cases may be explained in this way, but very often the pyramidal degeneration found in the cord does not extend to the medulla, and this was so in Köster's case where no degeneration was found at the level of the pyramidal decussation. Such cases seem to indicate that this degenerative process has its origin in the cord. Involvement of the meninges of the lateral region of the cord is not sufficiently invariable or intense to account for this process.

The columns of Clarke in the gray matter at the base of the posterior horns often present in cross-section pronounced evidence of involvement. This is not due in the main to implication of the cells of these columns but to atrophy of the end brushes of the fibers that enter through the posterior roots and terminate about the cells of these columns. The cells themselves for the most part escape and hence there is little or no degeneration of the direct cerebellar fasciculi that originate in these cells. Such degeneration however is not rare. In both of the cases in children there was degeneration of moderate degree in the terminal fibers of these columns and in Köster's case degeneration was found in the direct cerebellar fasciculi in the dorsal and lower cervical region but not above the latter level. Occasionally degenerative changes are to be found in the cells of the anterior horns. As a rule they are not extensive. In cases that have been characterized during life by localized muscular atrophy the changes in the anterior horns of the corresponding segments may be pronounced, but even such instances may be apparently due to involvement of the anterior roots, as Dejerine and Nonne and André Thomas have shown, rather than to degeneration of the cells. In this connection, as Nageotte and Schaffer have shown, the motor portion of the combined spinal roots shows a process similar to that found in the sensory portion, although much less marked. In cases, however, associated clinically with ordinary trophic disturbances, such as are common in tabes, Schaffer and Lapinsky have found marked degenerative changes in these cells, as well as in those of the central and postero-lateral gray matter. These findings have been confirmed by Etienne and Champy. It is probable that these changes were not found by earlier investigators on account of the methods of staining employed. Degeneration of white fibers running to and ending about the cells of the anterior gray matter is commonly found. These are probably reflex paths coming from the posterior roots. In the great majority of cases the process commences at the lumbar enlargement of the cord in the lumbo-sacral roots and the root involvement does not extend higher than the dorsal region. As the root fibers that at first form Burdach's columns in their further course upward to the medulla converge more and more toward the posterior median fissure and finally form the columns of Goll, it follows that sections through the cervical cord usually show complete sclerosis in the columns of Goll while the columns of Burdach at this level, and the corresponding posterior roots, present a normal picture.

Occasionally, however, the cervical roots show considerable implication, and now and then a case occurs in which the process commences in the cervical region and goes on to extensive and extreme degeneration. In this case the pathologic picture is somewhat altered, especially in cross-sections of the upper cord, for in this case the degeneration and sclerosis will be found in the columns of Burdach while the columns of Goll will be found uninvolved unless the process has appeared in the lumbo-sacral roots as well, in which case the posterior columns will show more or less complete sclerosis except for the cornucommissural tracts which invariably escape. However extensive the tabetic process may be, the first two or three pairs of cervical roots escape involvement.

The sclerosis of the posterior columns can be traced as far as the cuneate and gracile nuclei in the medulla.

The sclerosis of the pyramidal tracts often disappears by the time the medulla is reached and cannot be traced further. This was so in Köster's case, which is remarkable because the case was one of taboparesis. It is impossible at present to account for this. It was also true of the degeneration seen in the direct cerebellar tracts in the same case. It had disappeared in the upper cervical cord.

The degeneration in this case ascribed to Gower's tract was I think merely in the ventral spino-cerebellar fasciculus.

The tabetic process is approximately symmetrical although critical comparison of the two halves of the cord, will usually show slight differences in the degree or extent of involvement.

Tabetic Lesions Outside of the Cord Proper.—*Medulla, Pons and Basal Ganglia: Cranial Nerve Implication.*—Among the most common of tabetic lesions are degenerative changes in the cranial nerves and their nuclei. While almost no opportunity has been afforded for actual study of these lesions in childhood we may conclude, by comparison of the clinical symptoms in childhood with those of adult life, and basing our conclusions on the changes found in these adult instances, that such lesions are quite as common in the child as in later life. This is particularly true of involvement of the optic nerve. In fact if we may draw such inferences from the clinical symptoms it may be said that lesions of this nerve are more common in childhood and appear at an earlier period than among adult tabetics.

The process consists in a degeneration of the nerve fibers and a proliferation of the neuroglia. Opinions are divided as to the origin of the process, whether it commences in the retinal cells or whether the process is due to a sclerosis of vascular origin or to an interstitial neuritis, possibly secondary to meningitis, or whether it is simply a primary atrophy. The following is the result of the examination of the optic nerves in Köster's case in which the clinical symptoms, beginning with optic atrophy, appeared at about 12 and death occurred at 18: Complete loss of fibers in nerves, chiasm and tracts with increase of connective tissue, with over-filled vessels and signs of light grade of endarteritis (Köster). Complete degeneration of the ganglia cells of the retina with atrophy of the nerve-fiber layer, active proliferation of glia cells, especially about the papilla, no signs of inflammation except in the arteries which showed pronounced endarteritis, outer retinal layer normal, choroidal vessels free from change (Birch-Hirschfeld).

Degenerative processes in the oculo-motor nerves and in their nuclei, and the tracts connecting them, are very common, especially in the case of the third nerve. The ascending sensory root of the fifth is almost invariably involved, less frequently the motor portion of the fifth, nucleus and nerve. The nucleus of the tenth (nucleus ambiguus) the solitary bundle (descending roots of the ninth and tenth nerves), the nucleus of the twelfth, and finally the eighth nucleus may present degenerative changes. In fact no cranial nerve enjoys immunity from implication in the tabetic process.

Peripheral Nerves.—Extensive degeneration has been found in the peripheral nerves, especially by Oppenheim and Siemerling, Nonne, Dejerine and others. These changes have not been limited to the sensory filaments but have been pronounced in the intramuscular filaments of the motor nerves with amyotrophic changes. Degenerative changes have also been found in the terminal filaments about the joints in cases with joint changes.

Diagnosis.—It is difficult to conceive how the condition can be mistaken for any other disease. The only condition presenting a symptom-complex even remotely similar is multiple neuritis, and even this similarity is of the most superficial character. The form of multiple neuritis most common to childhood is that due to the toxin of diphtheria. Its conspicuous symptoms are ataxia for all finer movements, Romberg, staggering gait, loss of deep reflexes and muscular weakness. It develops rather acutely, within a few weeks of an inflammation of the throat, and ataxia is the earliest and throughout the course the dominant symptom, unless muscular weakness becomes so extreme as to render the child helpless. There is often a paretic condition of the palate, causing nasal voice. Optic atrophy never occurs, and pupillary and bladder disturbances are practically unknown, and sensory phenomena are either absent or of the most inconspicuous character. It is hardly necessary to say that crises, girdle sensations, etc., are not present. The course is moreover of but a few weeks duration and after its maximum is reached tends to steady improvement and complete recovery.

Syphilis, *limited to the cord,* is of extreme rarity in childhood, and when it occurs it usually produces pyramidal tract phenomena with the signs of spastic paraplegia, with exaggeration of the deep reflexes, and the presence of abnormal reflexes.

In tabes commencing with pronounced symptoms of paresis, there may well be for a time, some confusion with cerebral syphilis, but this will only be in cases where the paretic process persists as the dominant feature of the case, with the tabetic process but slightly developed.

Ataxic paraplegia can hardly be mistaken for tabes if it is borne in mind that it is a postero-lateral sclerosis, with pronounced spastic phenomena in the legs and that the pupils are never involved.

The confusion of tabes with cerebello-spinal ataxia (Friedreich-Marie) can only occur where there is utter misconception of the clinical characteristics of the latter disease. Unfortunately this misconception has existed and has led to the reporting of many cases as tabes which bear no resemblance to the latter affection. It is superfluous to compare the symptoms of the two diseases. Examination of the spinal fluid should never be neglected in any doubtful case. Whether or not the Wassermann test is negative, a cell count should always be made. A lymphocytosis of varying degree will be found, and will serve to exclude all other conditions with which the disease might be confounded with the exception of cerebrospinal syphilis and tuberculous meningitis. The cerebral symptoms sooner or later invariably present in both of these conditions in childhood will prevent error.

Prognosis.—The outlook is hopeless as far as cure is concerned. The course is slow and characterized by considerable periods during which little or no progress is made, while the phenomena as a whole present a somewhat milder picture than that seen in the average adult case. Nevertheless perhaps this is more apparent than real, on account of the relatively small number of cases observed, and the fact that only a few were under observation for long periods. Certainly the lightning pains and the crises may often be of great severity.

The immediate causes of death in adults are as follows:

Cardiovascular disease
- Cerebral thrombosis
- Bulbar paralysis
- Myocardial degeneration
- Pulmonary edema

Cardiorenal disease	Nephritis Uremia Pulmonary edema
Septicemia	Cystitis Pyelo-nephritis Decubitus
Terminal infections	Pneumonia Tuberculosis

Practically all of the cases commencing in childhood terminate in early adult life. The average duration in adults is from 10 to 12 years. As far as can be determined from the few cases in which the entire course is known, these figures correspond to the duration of cases commencing in childhood.

Treatment.—Since we have no data concerning the application of treatment in childhood as distinguished from that applied in adult cases, the latter must form the basis of measures to be recommended in childhood. There is no reason, however, to assume that the palliative measures applied in older tabetic cases should not be equally applicable to children.

Up to the time of the discovery of salvarsan the treatment resolved itself into the mitigation of symptoms as far as this was possible and the attempt to stop the progress of the disease by the methods employed for many years in combating syphilis of other parts of the body, namely, inunction of mercury or its intramuscular injection and the administration of mercury and the iodids by mouth. No curative results have ever been achieved by these procedures and could not reasonably have been expected in view of the destructive nature of the process and the impossibility of regeneration in nervous tissue so completely disintegrated as that in tabes.

Now and then the symptoms seemed to be lessened in intensity but sooner or later returned with their former severity, but more often little or no effects were to be observed.

An exception to this statement is to be made in those instances that presented symptoms of active syphilitic disease either within or without the nervous system. These symptoms often responded promptly to the usual anti-syphilitic remedies. Our efforts were therefore limited, in the average tabetic, to the amelioration of the patient's sufferings by the employment of opium and its derivatives, and to attempts to delay incapacity to the latest possible moment by the institution of exercises tending to lessen the disability caused by the disturbances of coördination.

The use of opium is attended by obvious and grave disadvantages and efforts have been made to find other drugs to lessen the pains. Of these the only one that has seemed to occasionally afford some relief in the writer's experience is aluminium chlorid in adult doses of 4 to 10 gr. in aqueous solution three times a day. Unfortunately this has often failed to secure relief. Whether the cases in which it seemed of some value were actually relieved by the preparation or whether the mitigation of pain was due to the intermissions often seen in these cases, I am unable to say. I have not seen much benefit from the ordinary analgesics—acetanilid, etc., when the pains have been severe, but they may be tried. The use of morphine during the crises is unavoidable. At times etherization may have to be resorted to. When the crises last for more than a day or two, rectal feeding should be employed. When Förster proved the feasibility of resection of the posterior sensory roots for the relief of spastic paralytic disorders it occurred to many clinicians that a similar operation might relieve the pains of tabes, which were known to be due to posterior root irritation. The first operation for this purpose was performed in New York, and within a few weeks the second was performed on a patient on our service at the Boston City Hospital. The relief in this case was immediate and lasted for six months, until the patient resumed excessive use of alcohol, when pain returned, and he soon after committed suicide. This man had been an intense sufferer for many years, and, like many in a similar predicament, had become a morphine addict. With the advent of salvarsan came new hope in the permanent achievement of therapeutic results in this as well as in other syphilitic affections of the central nervous system. It was soon found that salvarsan *per se* could not with safety be introduced into the spinal canal. In 1912 however, Swift and Ellis conceived the idea of administering salvarsan in the usual manner into the blood and very shortly afterwards withdrawing a quantity of blood and allowing it to stand, at a low temperature, until the corpuscles had separated from the serum, when this serum was injected into the spinal subdural space, replacing an equal quantity of cerebrospinal fluid withdrawn.

This method has now been employed on our service at the City Hospital in a large number of cases, and I am able to say definitely that it has resulted in amelioration of many of the subjective symptoms of this disease. In fact so convinced of their relief have the tabetics become that they return again and again for repetition of the procedure. Unfortunately the objective phenomena of the disease are not influenced in like degree, and I have not been able to convince myself that the process has been arrested. While we have now been employing this method for several years it is still too early to determine whether or not the progress of the disease is materially influenced. During the past two years we have been adding to the serum before injection a very small amount of diarsonal (salvarsan) which can be done with safety. While little or no change in the objective symptoms is to be expected, an exception is to be made in the case of the disability in gait and coördination. The improvement in this respect is sometimes quite strikingly obvious.

The details of the procedure at the City Hospital are in brief as follows, and I give the method that has been employed for children on my service. While these cases were not tabetic the technique is the same as would be indicated in tabes.*

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* This is the technique of Dr. George P. Sanborn.

From 0.1 to 0.15 gr. of salvarsan or diarsonal is injected into the blood in the usual manner. One-half hour later 40 to 50 c.c. of blood are withdrawn and allowed to stand in the sterile glass tube at room temperature for four hours, the clot is then separated from the sides of the tube by means of a sterile glass rod. When this rod is removed it is touched to a culture tube which is then placed in an incubator to be examined the following morning to determine the sterility of the blood serum before it is injected into the spinal canal. The blood serum in its tube is placed in an ordinary refrigerator and left there until the next morning. It is then removed from the tube and the separated clot, by means of a sterile pipette, and put into another sterile tube, the open end of which is then sealed in a blow-pipe flame. It is then put into a water-bath and kept at a temperature of 50 to 60°C. to inactivate or destroy the complement. If the culture is shown to be sterile, the serum, now amounting to from 20 to 25 c.c. is then taken from the sealed tube and injected into the spinal canal through the space between the third and fourth lumbar vertebræ. It is needless to remark that every step of the entire procedure should be conducted with the most rigid asepsis. The procedure should be repeated every 10 to 14 days, unless not well borne or unless, as may rarely happen, paralysis of the bladder occurs. Considerable headache occasionally follows the injection within 24 hours, but, even if severe, this need not cause alarm. The same may be said of the pains in the legs that not infrequently occur. The patient should be kept in bed until the following day in my opinion, although all are not so conservative as this.

In addition to these efforts to mitigate the severity of the symptoms, much can be accomplished by employment of the exercises first suggested by Fränkel for the relief of the ataxic disturbances. These consist of certain practice exercises designed to re-educate the nerve paths and centers in the transmission and control of coördinative impulses. The exercises have for their object the substitution of conscious directed muscular activity for the more or less subconscious automatic movements normally executed, but which have become more or less impossible in the victim of this disease. The technique will be found in the original publication.

Brief daily suspension of the patient in an apparatus, similar to that used in applying a plaster jacket, has its advocates and undoubtedly gives relief in some cases. It should be employed cautiously for not more than one-half to two minutes daily. Another method of stretching the lower cord is to place the patient on the floor with legs extended and held in that position while the head and body are gently bent forward, increasing the degree of flexion very gradually from day to day, and not continuing the procedure longer than two or three minutes at a time. Both methods may relieve the pains and the bladder disturbance.

Massage along the spine may also be employed with benefit, probably on account of its effect on the circulation and nutrition of the cord. It may also be used over any part of the body where sensory disturbance is troublesome, and gives relief to the subjective feeling of cold, often a source of discomfort to the patient especially in the feet.

Attention must be paid to the clothing for a similar reason, and where feasible a climatic change is advisable, especially in northern latitudes during the colder months.

Warm baths have also their value in lessening discomfort. Very hot or very cold baths should not be used.

Electricity has occasionally proved of value. The static or the high frequency current should be employed along the spine. Cystitis is very likely to occur and to add to the suffering of the later stages of the disease. It must be met by the usual measures which it is not necessary to deal with here.

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MYELITIS

If we except anterior poliomyelitis and the marginal myelitis that is often an accompaniment of meningeal inflammation, myelitis is a rare occurrence in childhood. Many conditions have been included under this title that belong in an entirely different category. This has resulted in much confusion in the comprehension of the term. Softening secondary to vascular accidents or disease should no more be included under this classification than cerebral hemorrhage or thrombosis under the classification of encephalitis. It is conceded that a hemorrhage into either the brain or cord is followed by a local inflammatory reaction, and that this reaction is an encephalitis or a myelitis as the case may be, but this is a purely circumscribed condition and not a clinical entity in any sense. We know that a disease exists that possesses all of the clinical and pathologic characteristics of an inflammation of the cord, just as we know that such a disease as encephalitis exists, and to that disease the term myelitis is here applied and in this sense the term is employed.

Myelitis is entitled to rank as a disease *sui generis*. It may be either acute or chronic. The latter form is rare in adults, unknown in childhood.

ACUTE MYELITIS

Acute myelitis may be focal, disseminated, or transverse, and it may affect the gray or the white matter, or both, and the clinical picture presented by any given case naturally depends upon its location and extent.

Etiology.—The disease has all of the characteristics of an acute infection. Whether the infection acts directly by bacterial invasion of the cord or whether it acts indirectly by means of toxic products is not clear in the light of our present knowledge. Logical deduction, and reasoning by analogy from our knowledge of meningitis and encephalitis, seem to support the probability that it is always the result of actual bacterial invasion. A strong support to this hypothesis is furnished by the extreme frequency of intense toxemic conditions, due to bacterial growth elsewhere in the body, and the relative rarity of acute myelitis. Various organisms have been found in the cerebrospinal fluid. The most commonly found have been the common pyogenic organisms—staphylococci and streptococci—but pneumococci and typhoid bacilli have occasionally been demonstrated. It is quite probable that these organisms were not the actual cause of the myelitis. While it cannot yet be said that these pyogenic organisms may not cause a typical myelitis, it can at least be said that the disease does occur without the presence in the cerebrospinal fluid of any organisms, discoverable by known methods of staining or by ordinary microscopic technique. The analogy between this fact and the long search for the organism responsible for anterior poliomyelitis suggests that the real cause of other types of myelitis has not yet been discovered.

As we shall see later we do not know that the organism responsible for anterior poliomyelitis may produce a more diffuse myelitis than the type usually seen. Certain of the acute ascending cases seen during an epidemic of poliomyelitis are unquestionably caused by the same organism.

The disease has seemed sometimes to follow the acute general infections and cases have been reported following measles, diphtheria, typhoid, scarlet fever, influenza and pertussis. Some of these instances are questionable cases of myelitis, and more probably of cerebral origin. Myelitis may occur at any age. The acute transverse type cannot be said to show a predilection for any particular period of childhood, a rather striking contrast to anterior poliomyelitis, in this respect.

Symptomatology.—Among the infrequent cases occurring in childhood that known as acute transverse myelitis and the acute ascending form, known as Landry's paralysis, are the only types requiring clinical description.

Acute transverse myelitis in childhood, like acute anterior poliomyelitis, is characterized by the relative insignificance of premonitory symptoms. It may in fact occur without either subjective or objective warning, and the paralysis may be the first sign of the existence of a serious disease of the cord, appearing as suddenly as an apoplectic attack, or the victim may retire apparently in good health and be found with paralyzed legs in the morning. On the other hand, while the onset is almost always abrupt, the height of the paralysis is not always so quickly attained. The child may fall in the midst of its play, complain of pain in limbs, back or head, be put to bed, and subsequently be found to be paralyzed in the legs. Fever may or may not be present. If it occurs it does not remain elevated as a rule. For this reason it is perhaps overlooked and often thought to be absent when it may have occurred at the onset.

Retention of urine is almost invariably present and persistent, requiring relief by catheterization. It is an initial symptom as is also obstinate constipation. In addition to the initial pains, when they occur, paresthesia is sometimes complained of but often not clearly described by the child. These sensory subjective phenomena may appear in one extremity some time before appearing in the other. This is also true of the paralytic symptoms, although the interval at the most is but a few hours. There occur occasional exceptions to this rule and in such cases several days may intervene between the appearance of the symptoms in one extremity and their appearance in the other.

Some cephalic retraction and a considerable degree of cervical rigidity may be among the early symptoms, and they may persist for some days. Vomiting within the first day or two may occur, but it is not a conspicuous initial symptom and usually follows attempts to administer a cathartic, such as castor oil. Convulsive attacks are decidedly uncommon but they occasionally appear, not as an initial symptom but during the early days of the disease. Consciousness may not be lost, as in a case of the writer's where clonic movements of all four extremities occurred, although the child was perfectly conscious and asked for water.

Where pain and paresthesic sensations constitute the only premonitory symptoms the child is usually allowed up and about and, although it may give evidence of the approaching collapse in the legs by frequent falls, but little is thought of this at the time. The child goes to bed and in the morning when it tries to stand falls helpless to the floor, or where the ileo-psoas and back muscles are involved it may be unable to sit up in bed.

This is the paralytic stage and the paralysis at this early period may be either flaccid or spastic, but the latter is the rule. It is almost always limited to the legs. With this paralysis, and corresponding quite closely with it in conformity with the segment lesion of the cord, there will be found usually some degree of sensory impairment, not often an absolute loss so much as a diminution, usually involving all forms of common sensation, but occasionally, especially when the case is seen later, showing some discrimination between the forms. A hyperesthetic zone may occasionally be found above the level of hypesthesia, during the acute stage. During this paralytic stage, if the type of the paralysis is spastic, there is obstinate retention of urine, although there may seem to be incontinence from dribbling from the overfilled bladder, or flooding of bed or clothes from sudden involuntary giving away of the sphincter in response to the tension within the bladder. The bowels are constipated, but may be spontaneously evacuated without the consent or even the cognizance of the patient.

If sensation is seriously impaired bedsores may develop, usually on the buttocks and over the malleoli, and they may occur even in the absence of sensory impairment where confinement to bed is prolonged, as it usually is.

It is obvious that the particular type of disability produced by the disease depends upon the particular level of the cord affected and the extent of destruction in a transverse direction. The disease shows a predominant predilection for the dorsal region of the cord, in the child as in the adult, and, however complete the picture of a transverse lesion of the cord may be, shortly after its onset, this completeness, if it is at first present, tends to recession, as time goes on, leaving evidence of varying degrees of permanent injury to the pyramidal tracts, and, to a less extent, not infrequently, to the sensory tracts as well, or more rarely, to some circumscribed area of the gray matter of the cord.

Lumbar puncture may show a perfectly clear fluid, not under pressure and without material increase of its cellular content. When lymphocytosis occurs it is always moderate. Polynucleocytosis does not occur unless there is an invasion of some of the common pyogenic organisms. Globulin may be positive.

After the symptoms have become fully developed there ensues a stationary period, often of long duration, during which little or no improvement is noticeable.

The course of the disease is well illustrated by the following cases:

Boy four years old. While at play he fell down and when picked up he complained vaguely of legs and back. He was somewhat feverish and complained of pain in the head and abdomen. The head was somewhat retracted and the neck stiff. Two weeks after the onset of these symptoms he had a convulsion with twitching of all four extremities, lasting several minutes. This recurred once some days later. When placed on his legs he was found to be unable to stand. He presented extreme spastic paraplegia, when seen six months later, and a diminution of tactile perception that extended over the entire surface of the lower extremities and over the trunk to the level of the navel. There was lack of control over the urine and feces. He was unable to stand. There was great exaggeration of the deep tendon reflexes in the legs and double ankle clonus. When again seen, at the age of 12, his condition was similar except that he could walk and had been able to do so fairly well since he was five years old, although he fell frequently whenever he tried to hurry.

He showed marked upper dorsal scoliosis, convexity to the right, dorso-lumbar convexity to the left. He wet the bed nightly and occasionally wet his clothes and sometimes soiled them when unable to reach the toilet promptly.

Boy 12 years old. Previously perfectly well until his eleventh year, when he had diphtheria, from which he made a good recovery. When 12 years old, he complained of pains in both hips and of paresthesic sensations in both legs. These continued for four or five days and were considered "growing pains" by the medical attendant. Then one morning when he got out of bed he found that he could not bend forward, and when he attempted to walk across the floor he fell. He nevertheless kept about that day, but at night lost control of his right leg, and on the following day lost control of the left leg. The upper extremities were unaffected. There was no vomiting or any apparent febrile disturbance. He was constipated and unable to pass urine spontaneously. He presented a spastic paraplegia, when seen some time after the onset of the trouble, together with considerable atrophy of the right glutei and posterior muscles of the right thigh. He developed bedsores on the right buttock and over both internal malleoli, during two years of helplessness. At 15 years there was some retardation in growth of the right leg and a scoliosis. He could only walk with crutches.

In this case, we see, as the residual of a transverse myelitis, bilateral permanent injury to the pyramidal tracts and partial unilateral destruction of anterior horn cells in the lumbar cord.

Boy five years, four months old. No previous disease. Vaccinated two weeks ago. Three days ago, without premonitory symptoms, sudden retention of urine, constipation and inability to walk. On the following morning he vomited. No headache and no apparent fever. He required catheterization and enemata.

When seen on the third day, there was some cervical rigidity and double Kernig sign. Upper extremities were normal.

Below the cervical region there was no spinal rigidity but passive flexion and extension caused abdominal pain. Abdominal reflexes were abolished, cremasters sluggish.

There was a total loss of voluntary movement in both legs and in the ileo-psoas on both sides. Plantar irritation caused some involuntary movement in the flexors of thighs and legs.

Deep tendon reflexes in legs were increased. There was no ankle clonus or Babinski at this time.

Below the sixth dorsal level there was diminished tactile and pain perception, somewhat more marked on the left side. Temperature 99 degrees; P. 105; R. 23; urine negative.

Spinal fluid (fifth day of the disease) crystal clear, under no hypertension, 25 cells per cubic millimeter, all lymphocytes. No red cells.

Blood showed leukocytes: 77 per cent. polynuclear, 23 per cent. mononuclear.

The rigidity of the cervical muscles disappeared on the fourth day, and the sensory disturbance had receded on the fifth day to the level of the iliac crests, and gradually disappeared entirely.

The temperature varied between 99 and 101.3 degrees for several weeks.

Voluntary movement gradually returned during the following month, leaving a typical spastic paraplegia of permanent character.

Pathology.—The process consists of a diffuse segmental inflammation of the gray and white matter of the cord. The character of the process strongly suggests that the entry of the infectious agent is through the blood-vessels. There is pronounced perivascular infiltration with round cells, often accompanied by small hemorrhages and by thrombosis of the smaller spinal vessels. With this process there occurs a disintegration and disappearance of the parenchymatous elements of the nervous structure with a secondary proliferation of the interstitial elements. The meninges may be implicated locally to some extent. Indeed the clinical symptoms, such as cervical rigidity and cephalic retraction, would suggest that such implication is possible some distance from the site of the myelitis.

The segment or segments involved are softened, presenting a "collapsed" or flabby consistence, usually of a faintly yellowish hue, but if seen in the earliest stage of the process a slight pinkish tinge may be apparent. Eventually the section is of a grayish hue and shrunken appearance. On cross-section the gray and white matter is poorly differentiated, but this depends upon the extent to which the gray matter may have suffered in different instances.

Diagnosis.—In childhood at least the diagnosis is not difficult. Only when trauma has occurred shortly preceding the onset of symptoms may the diagnosis remain long in doubt. Under such circumstances, especially if the trauma has been such as to involve the spine, differentiation between transverse myelitis and hematomyelia may be impossible. From a practical standpoint the differentiation is of little importance for the ultimate outcome is equally unfavorable. Great care, however, must be observed under these circumstances not to overlook a compression of the cord from vertebral dislocation or fracture, a condition capable of relief and sometimes complete restitution.

It must also be kept in mind that the symptoms of vertebral caries may appear suddenly, especially the cervical form, when the condition has been entirely unsuspected, and produce symptoms, from transverse compression of the cord, exactly similar to those caused by transverse myelitis.

In both of the latter conditions a radiograph will often clear the doubt, but even without its aid, care will usually prevent error. The discovery of spinal deformity, even of slight degree, will at least turn the thought to either of these possibilities. Examination will disclose, under these circumstances, well marked but localized muscular spasm in the spinal muscles in proximity to the bony lesion whenever attempts are made to passively flex or extend the spine, accompanied by complaint of intense pain. Pain may also be elicited by firm pressure downward on the top of the head.

For a brief period certain instances of anterior poliomyelitis may simulate acute transverse myelitis. This is not surprising when we reflect that we are simply dealing with a type of myelitis the ultimate effects of which are limited to the anterior gray matter but which in its acute stage may consist of a diffuse myelitis. Nevertheless there is almost always an asymmetry, even in such instances, in the distribution of the paralysis, the latter is moreover always of the lower motor neurone type, and the pyramidal fasciculi invariably escape, and there is usually an entire absence of sensory symptoms. Moreover involvement of the arms is rare in transverse myelitis while extremely common in anterior poliomyelitis. Transverse myelitis in childhood is rarely so intense as to present a picture of complete lower segment paralysis below the level of the lesion. If it does this the abolition of sensory function as well is so striking as to immediately rule out anterior poliomyelitis.

Multiple neuritis always presents the picture of lower segment paralysis, never any signs of pyramidal implication. Moreover it is gradual in development and the paralysis shows a tendency to select certain muscle groups, such as the extensors, affecting them chiefly or exclusively. In the form most frequently seen in childhood, that secondary to diphtheria, disturbances of coördination, station and gait occupy the foreground of the clinical picture, and could not possibly be mistaken for myelitis. Finally sphincteric disturbances are absent in multiple neuritis. The only other condition likely to offer difficulties in diagnosis is so rare in childhood as to be a negligible factor. This is vascular disease of the spinal vessels. The only factor causing such disease in childhood is syphilis. In syphilitic arteritis thrombosis may produce symptoms of typical segment lesion of the cord, including spastic paraplegia. In the very rare instances where spinal symptoms precede the cerebral phenomena the differentiation between the two conditions can be made only by a complete examination of the spinal fluid or by the therapeutic test of intensive antispecific treatment.

Very rarely latent and unsuspected tumors of the cord may simulate myelitis, when phenomena due to edema or to hemorrhage suddenly make their appearance and produce a picture more or less typical of a transverse lesion. Such cases may remain for a time doubtful but they do not long remain stationary, as in myelitis, and sooner or later the progress of the tumor-growth reveals the cause of the clinical phenomena.

Prognosis.—During the active stage the prognosis is always doubtful, for it is impossible to tell where the process is going to stop. As a matter of fact, however, by the time the diagnosis is made the height of the process has usually been reached and the limitations of the lesion fairly evident. Only occasionally do subsequent exacerbations of the process alter the clinical picture or modify the prognosis. Whenever the symptoms point to involvement of the cervical cord the prognosis becomes grave, through danger of implication of the phrenic nucleus, at the fourth cervical segment. As a rule the predilection for the dorsal or lumbar cord makes the prognosis as to life good. It is hopeless, however, as far as *restitutio ad integrum* is concerned. It is possible that cases have entirely recovered, but in such instances the diagnosis is to be questioned. Improvement, however, is sometimes greater than would be anticipated from the degree of helplessness seen shortly after the onset. Like other cord diseases, cystitis and bedsores are troublesome factors, and may give rise to sepsis. Fortunately the usual spastic palsy remaining as a residual of the process does not permanently confine the victim to bed, and the bladder disturbance is eventually not such as to lead to cystitis provided this has been escaped during the necessary catheterization of the early period. The sensory disturbance also does not remain permanently so severe that bedsores are to be feared as a chronic complication.

The greater the restriction of activity the more susceptible the child becomes to tuberculous disease, but this is true of all conditions that similarly restrict exercise and its concomitants, proper respiration and circulation.

Treatment.—Usually there is insufficient warning to give opportunity for direct treatment even if we possessed a specific therapy for the disease, which of course is not the case. Our efforts must be confined to the mitigation of the immediate symptoms, to the prevention of complications during the early period of helplessness, and eventually to the overcoming, so far as possible, of the disability incident to the paralysis.

There is but little need of treatment during the onset so far as subjective symptoms are concerned. Occasionally sedatives may be required. Where headache is troublesome an ice-bag or Leiter's coil may be used.

Ice-bags to the spine are a source of more discomfort than any theoretical benefit possibly to be achieved by their use.

Retention of urine is invariably present and should always be kept in mind, even when not complained of.

The catheter will usually have to be used and it must be employed under the most rigid aseptic precautions. It is always well to administer hexamethylenamin as long as catheterization is necessary.

During the long confinement to bed the most solicitous care of the skin, especially over the sacrum, buttocks, trochanters and malleoli, must be observed. Absolute cleanliness and dryness must be maintained, and pressure on any one area of skin relieved by frequent changes of position and by air cushions or by an air mattress.

Contractures should be anticipated and avoided as far as possible by a cradle to prevent the weight of bed-clothes resting on the feet, or by placing the legs and feet in apparatus that will prevent drop-foot or the position of equinus, and therefore lessen the likelihood of extreme contracture of the calf muscles. The same care must be observed in the case of the adductors of the thighs and the hamstring groups.

As soon as possible massage, passive stretching and educative exercises should be instituted and continued for months.

Absolute rest for the first few weeks and then efforts to get the child about as soon as possible should be the rule of treatment in these cases as in ordinary anterior poliomyelitis. Orthopedic measures are indicated in the same measure that they are indicated in the latter disease, but especially tenotomy of the spastic contractured muscles. In cases with extreme flaccid paralysis that does not improve after several months of treatment braces may assist to get the child about and afford it necessary exercise.

ACUTE ASCENDING MYELITIS
(Acute Ascending Paralysis—Landry's Paralysis)

An acute infectious inflammation of the cord, chiefly affecting, but not confined to, the gray matter, usually commencing in the lower portion of the cord and advancing upward as far as the medulla.

Etiology.—The etiologic factor is unknown. It is of interest however that, whereas isolated instances of the affection are so infrequent as to make the disease a rare one, numerous cases have been met with, during the severe epidemic of anterior poliomyelitis of recent years, that correspond in every essential particular with the acute ascending paralysis described by Landry. It is extremely probable that the disease is caused by a highly virulent strain of the organism responsible for anterior poliomyelitis. It is certainly not caused by any other known variety of organism. The same multiplicity of organisms have been reported in this affection as were formerly thought to be of importance in the etiology of the more common disease, and quite as frequently the search for organisms has been without results.

Symptomatology.—Like other forms of myelitis, the disease may commence with or without prodromata. If the latter are present they consist simply of the symptoms of acute toxemia, general malaise, light febrile disturbance, loss of appetite and occasionally headache and vomiting. Often there are no prodromata of general character but the disease begins with a subjective feeling of numbness in the feet rapidly succeeded by weakness which very quickly resolves itself into a rapidly ascending flaccid paralysis with total abolition of reflexes. There is rarely any pain, and even paresthesia may be entirely lacking. Notwithstanding the completeness of the lower segment paralysis, the sphincters are not involved, although there are a few exceptions to the rule.

The paralysis rapidly advances to the trunk. Here there may seem to occur a slight delay in advance but not for long, and very soon the abdominal muscles, the lower spinal muscles, and then the intercostals are paralyzed and respiration becomes mainly diaphragmatic, with still some help from the upper accessory muscles of respiration for a brief period. The arms are next involved and the muscles of the cervico-dorsal spine. Death now usually ensues from destruction of the phrenic center at the fourth cervical segment. If this is delayed, medullary phenomena, in the form of disturbances of speech and deglutition and failure of cardiac function, occur.

The facial and ocular muscles may also be implicated although death usually occurs before their nuclear level is reached. The sensorium remains clear to the end, except in so far as it may be affected by deficient oxidation of the blood in the last few hours, from the respiratory failure.

Very rarely the paralysis commences in the arms and, after ascending to the shoulders, descends throughout the rest of the body.

Pathology.—Like other types of myelitis there is evidence of vascular and perivascular inflammation, with acute parenchymatous degeneration in the cells and axones of the gray matter of the cord. The process, however, does not stop here and cases have been reported with widespread changes in the peripheral nerves of inflammatory character. A few cases have been included under the head of this disease, where peripheral neuritis was the only change found. Such instances have no place here.

That the disease is an acute infection is abundantly demonstrated by those cases in which pronounced changes in spleen, liver, kidneys and lymph-nodes have been found.

Diagnosis.—There is no other condition likely to be mistaken for this disease, if we include under Landry's paralysis those instances seen during epidemics of anterior poliomyelitis. Until it is proven that the etiology is a different one, it seems to the writer that any form of acute rapidly ascending myelitis should be so classified.

The symptoms of acute multiple neuritis may simulate this disease in those rare instances of very rapid spread of the paralysis. It is conceivable that under such circumstances a differential diagnosis might be impossible if subjective and objective disturbances of sensation were absent.

Prognosis.—Usually hopeless. Very rarely, after reaching a certain height, the process may recede. One can never be sure of this, however, for after a certain degree of recession it may again advance to a fatal termination.

Death from respiratory paralysis usually occurs within a week of the onset, sometimes in two or three days or even sooner in cases commencing with bulbar signs. In cases that recover convalescence is prolonged, and recovery from the paralysis is incomplete.

Treatment.—Beyond supportive measures, such as stimulation, and such measures as are indicated in any toxic condition, free elimination, especially from the intestinal tract, there are no measures to be recommended as having any influence on the progress of the disease. Repeated lumbar puncture will apparently do no harm and would seem in fact a rational procedure on theoretical grounds, but there is but little hope of its delaying the advance of the process within the cord itself.

When the etiology is finally determined, the first hope of efficient therapeusis will be founded on scientific grounds.

DISEASES OF THE SPINAL MENINGES

External Pachymeningitis.—Inflammation of the outer layer of the dura is always secondary to some other process, chiefly such as involve the neighboring bone, tuberculosis of the vertebræ being the most common cause in childhood. Nearby suppurative processes may, however, produce an inflammatory reaction in the dura even when the bone is not apparently involved. It has been found in purulent inflammation of the pleura and peritoneum, and even in cases that have suffered with bedsores. This layer of the dura reacts also to the irritative effect of neighboring tumors.

While these primary processes may affect exclusively the external layers of the dura, there is of course no necessary limitation, and more often than otherwise the entire dura is implicated, and even the pia-arachnoid and the underlying cord.

Symptomatology.—The early phenomena are those indicative of spinal root irritation, pain and paresthesia, less often anterior motor-root irritation; later phenomena are those of transverse lesion of the cord, not usually complete although it may become so, but chiefly bilateral interference with pyramidal conduction—spasticity in the legs with increased deep reflexes and the presence of abnormal reflexes.*

Pathology.—The dura is thickened and sometimes covered with purulent deposit, when the primary source of the process is purulent. When the dura is greatly thickened the cord itself may be compressed, but it is pretty well established that the cord symptoms, when present, are not necessarily due to pressure but are often the result of secondary inflammatory reaction.

Prognosis.—This depends entirely on the character of the exciting cause.

Treatment.—This also resolves itself in the treatment of the primary factor.

Internal Pachymeningitis.—Pachymeningitis cervicalis hypertrophica. This disease, the first description of which we owe to Charcot and Joffroy, may be confined to the cervical region of the cord, but it is sometimes of greater extent, and there is often coincident involvement of the cerebral dura, as is well illustrated by Probst's† case in a 15 year old girl, in whom the condition was found in the cervical and dorsal cord and in the cerebral dura as well.

Starr believes that he has seen instances in which the clinical symptoms indicated a similar process in the lumbar cord.

Etiology.—The etiologic factors in the causation of the process are unknown. While a similar condition occurs occasionally in cerebrospinal syphilis and in general paresis it is not established that all instances have a similar etiology.

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* See physiology of the cord and symptoms of pyramidal tract lesions.

† Probst, Ueber Pachymeningitis, etc., Archiv fur Psychiatrie, iii, Bd. 36, (1902), p. 114.

Symptomatology.—The symptoms are those of compression of the upper cord, and the earliest symptom is pain and stiffness in the back of the neck. The pain eventually radiates, through the particular roots that happened to be compressed, down through the branches of the brachial plexus in the arms. These pains have all the characteristics of root irritation and may be associated with paresthesia, or eventually with anesthesia when the condition reaches the stage of paralysis. The process is chronic, and in the course of a few months there ensues the stage of paralysis. This paralysis first appears in the form of a gradually increasing spastic paraplegia, indicating the gradually increasing interference with transmission of impulses through the pyramidal tracts at the site of the disease. As the thickening progresses the gray matter of the anterior horns becomes implicated by a compression myelitis, or the anterior roots are directly involved, and a lower segment type of paralysis occurs, flaccid paralysis, with atrophy and the usual features of lower segment lesion, corresponding to the level of the disease. This flaccid paralysis however may not appear and the clinical picture may remain that of spastic paralysis in the legs.

The particular group of muscles implicated in the flaccid paralysis, when it occurs, depends merely upon the particular level of the cervical cord at which the anterior cells or roots are implicated. The extent of this type of paralysis is no index of the process, but merely shows that at that one level the pressure has been sufficiently severe to interfere with the function of these neurones.

During the stage of onset of the spastic phenomena clonic twitchings may appear in the muscles subsequently the seat of spasticity.

During the periods of hemorrhagic exacerbation the spinal fluid usually contains blood.

Pathology.—The process consists of a fibrinous exudate on the internal surface of the dura. In its earliest stage this consists of a very thin transparent membranous exudate. Into this pseudomembrane blood-vessels penetrate from the vascular inner surface of the dura and rapidly multiply. These newly formed vessels, like those in ordinary granulation tissue, are extremely thin-walled and overfilled. Hemorrhage therefore easily occurs, the hemorrhagic exudate in turn is organized and permeated by equally fragile vessels, which in their turn rupture, and so the process of building up layer after layer of fibrous membrane goes on, the older layers becoming inseparably fused with the dura, although sometimes the various older layers are not so closely bound together and are distinguishable.

Diagnosis.—This is impossible in the early stages of the process before the thickening becomes sufficiently advanced to interfere with the spinal function. Even then the condition cannot well be differentiated from spinal tumor or abscess, or even from transverse myelitis when, as may happen, symptoms of transverse compression appear rather rapidly. This process in the cord is sometimes associated with a similar process in the brain. In this case cerebral symptoms may appear and so dominate the scene that the cord lesion may be entirely overlooked and discovered only at autopsy.

The affection can be diagnosed only when the symptoms are purely spinal and all other possible conditions may be eliminated, and when the clinical phenomena show exacerbations, separated by distinct intervals during which the symptoms are stationary. If blood is found in the spinal fluid during the exacerbations, the presumptive diagnosis is greatly strengthened.

Prognosis.—The affection is usually fatal, death occurring during some one of the exacerbations. Many exacerbations may occur however before the fatal termination takes place. What proportion of cases, if any, recover is not capable of demonstration on account of the difficulties of diagnosis.

Treatment.—No treatment is known to have any effect on the process except in such instances as are of syphilitic origin. Then an intensive course of mercury and iodid is usually effective, or arsenobenzol may be used intravenously, or intraspinal treatment by the Swift-Ellis method may be employed.*

Spinal Leptomeningitis.—Except in local involvement of the pia-arachnoid, due to circumscribed neighboring disease, the involvement of the leptomeninges of the cord is practically always but part of a widespread leptomeningitis, in which the involvement of the cerebral leptomeninges is the primary and the predominant process, both pathologically and clinically.

In these instances the spinal process may be practically limited to the cervical region of the cord, although more often merely more intense there than elsewhere.

Under these circumstances the symptomatology is that of cerebrospinal meningitis and the clinical picture is that of a cerebral process, and is therefore not to be here described. While uncommon, cases do occur in which the spinal process is the dominant feature and these cases only are to be here considered.

Etiology.—These cases are of infectious origin. The organism may be that of ordinary cerebrospinal meningitis, or some one of the other common varieties of organisms, pneumococcus, staphylococcus, streptococcus, etc. Phenomena of the spinal type of meningitis are extremely common in acute anterior poliomyelitis, and a certain proportion of these cases belong in that category, especially such as present the sequelæ of flaccid paralysis.

It is sometimes seen as part of a general sepsis. It may follow vertebral fracture, penetrating wounds of the spine, or occur subsequent to operations on the spine, especially when the dural cavity has been opened. Several cases have been reported of tuberculous leptomeningitis limited to the spinal meninges, but this is not a common occurrence. Even when previous tuberculous disease of the vertebræ has existed, if the tuberculous process involves the meninges beyond the immediate site of the vertebral involvement, a cerebrospinal tuberculous meningitis usually results. In my experience also syphilitic meningitis limited to the spinal meninges has been very rare in childhood, although not rare in the adult.

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* See treatment of tabes, for details of this treatment.

Symptomatology.—As in cerebrospinal meningitis, the acute forms of spinal leptomeningitis in childhood may commence with headache and vomiting and even convulsions, especially in young children.

In cases of less acute onset there may be a brief period of general malaise. Rather obstinate constipation is the rule.

Early cervical rigidity is almost invariably present, attended usually by a certain amount of cephalic retraction.

All of these early phenomena are equally characteristic of cerebrospinal meningitis. Only in the later phenomena is it possible to detect the predominant or exclusive implication of the spinal meninges. After a somewhat variable period, usually but a few days, the phenomena become distinctly of spinal type, and indicative of root-irritation, hyperesthesia, pain and muscular spasm. Considerable pain in the back is sometimes complained of. Abdominal retraction, tâche, Brudzinsky's and Kernig's signs are usually present.

The temperature is considerably elevated, especially in the early stage, but presents no special characteristics.

Neither pulse nor respiration show the variations and lack of correspondence with the temperature so frequent in cerebral meningitis. If such occur, it is evident that the inflammatory process is no longer confined to the spinal meninges but that a cerebral basal meningitis exists.

In these spinal cases flexor spasm in the extremities, especially in the wrists and fingers, with subsequent contractures, is common, particularly in instances that run a subacute course. These are usually cases that commence with symptoms of cerebrospinal meningitis and, after a variable period of acute phenomena, subside into a subacute chronic course, in which the spinal symptoms predominate.

In cases that do not terminate fatally within a few days, the clinical picture may eventually be that of spastic contracture, as just described, or flaccid paralysis in one or more muscle-groups may result. We used to designate the latter cases as "flaccid paralysis following meningitis." It is probable however that these were instances of anterior poliomyelitis, with well pronounced meningeal involvement, and they are now so classified.

The majority of cases of leptomeningitis are accompanied by more or less marginal myelitis. If this myelitis remains peripheral no cord symptoms, beyond those of root-irritation, may occur. If the myelitis extends more deeply into the cord indications of interference with the cord functions results. This is more apt to occur in the postero-lateral field than in the anterior, and hence spastic phenomena are more commonly seen than flaccid conditions, owing to the implication of the pyramidal fasciculi.

The injury to the underlying cord substance is also furthered by vascular thrombosis or by secondary hemorrhages, both of which occurrences are common in all forms of meningitis. Syphilitic spinal lepomeningitis is characterized clinically by spastic phenomena, a slowly increasing rigidity in the lower extremities, with exaggeration of the deep reflexes and the appearance of other phenomena of pyramidal tract disturbance, the Babinski and Oppenheim phenomena and ankle clonus, also difficulty in emptying the bladder may occur from spasm in the vesical sphincter.

Pathology.—With the exception of the tuberculous and syphilitic varieties of spinal meningeal involvement, spinal leptomeningitis is usually purulent, consequently the pia is usually found covered with purulent or fibrino-purulent exudate. This may be more marked in some cases than in others, and is almost invariably more pronounced in the posterior than in the anterior meningeal field.

The pia loses its translucent appearance and becomes cloudy and more or less opaque. Close inspection reveals the fact that the process is more pronounced along the course of the blood-vessels, and this is confirmed by microscopic examination. Intense perivascular infiltration is found in the vessels of the pia and about their branches within the cord. With the intense congestion in these vessels areal capillary hemorrhages are associated.

The implication of the nerve-root is shown by disappearance of the myelin sheath in many of the fibers, with swelling of the axis cylinders and secondary degeneration, and similar changes are found in the white fibers of the cord, adjacent to the inflamed meninges, and penetrated by their vessels. This destruction of the cord substance proper is usually of marginal character but may extend more deeply into the substance of the cord. Secondary degeneration along the axis cylinders occurs.

The destructive process is accompanied by proliferation of the neurolgia and connective tissue. Thus secondary areal sclerosis may occur here and there throughout the cord, or the sclerosis may assume a more systemic character, especially along the pyramidal tracts. Adhesions between the pia and the dura are extremely common.

Diagnosis.—The diagnosis offers no special difficulties so far as the existence of an inflammation of the spinal meninges is concerned. The only difficulty that arises sometimes is the determination of whether the process is primarily a meningitis or an acute myelitis with incidental meningeal involvement. From the symptoms alone in the early stage it is often impossible to determine which is the dominant process. Under these circumstances lumbar puncture is of paramount value.

The great majority of instances of spinal meningitis in the strict sense of the term, that is where the process is primary in the meninges are caused by pyogenic organisms, and the cerebrospinal fluid is cloudy and contains an excess of polynuclear leukocytes.

In acute anterior poliomyelitis the fluid as a rule is clear and contains a moderate number of lymphocytes, with few if any polynuclear leukocytes. In acute transverse or acute diffuse myelitis the fluid is invariably clear and its cellular content is either normal or contains a slight increase in the number of lymphocytes. In myelitis the paralytic phenomena are predominant and usually early symptoms, and pain is less conspicuous. All of these affections are often accompanied by a considerable degree of meningeal involvement in the stage of onset, and under these circumstances differential diagnosis may be impossible and only the subsequent course of the case will reveal the true condition. This is especially true of anterior poliomyelitis. The early loss of the reflexes and the relaxed condition of the extremities, even before actual paralysis has appeared, together with the fact that these cases usually appear during an epidemic of the disease, will usually give the clue to the nature of the trouble, in instances where lumbar puncture is not possible.

Tuberculous meningitis limited to the cord is of such rarity as to be hardly considered. In cases of tuberculous disease of the vertebræ the appearance of signs of local or general meningeal irritation will of course suggest the dissemination of tubercles in the meninges. Except where the process is very circumscribed, the spinal fluid shows lymphocytosis, usually of pronounced degree.

Syphilitic spinal meningitis is always of gradual onset and the symptoms of root-irritation, so prominent a feature of acute meningitis, are usually lacking. The spinal fluid, as in tuberculous meningitis, shows a lymphocytosis but as a rule of much less pronounced degree. The Wassermann reaction of the fluid is of course a determining factor in cases of doubt. It should always be borne in mind that a negative reaction in the blood does not imply a negative reaction in the cerebrospinal fluid, and a test of the latter should always be made where doubt exists.

Prognosis.—The outcome of acute spinal meningitis depends entirely on the virulence of the infecting organism. The form of which we are speaking, namely that limited to the spinal meninges, is somewhat more favorable than diffuse cerebrospinal meningitis. During the acute stage the prognosis is always grave however, and only too frequently the process extends to the cerebral meninges, especially at the base, and the prognosis becomes that of general meningitis.

If caused by the meningococcus the prognosis is of course favorably modified by the possibility of the use of anti-serum.

Treatment.—For other acute forms but little can be done. Frequent lumbar puncture however may relieve the irritative phenomena and may possibly favor the outcome by diminishing the concentration of toxins in the cerebrospinal fluid.

Active catharsis should be employed in the stage of onset. Later sedatives are required for the relief of motor restlessness, spasm and pain. Ice-bags to the spine are not well tolerated by children, and are of doubtful efficiency.

Counterirritation is of little value and is not without danger in cases that later assume a chronic or subacute course on account of the tendency in such cases to develop decubitus. As in cerebrospinal meningitis, instances occasionally occur in which the fluid is clear, as in tuberculous meningitis, but without the lymphocytosis of that disease, and with a negative bacterial examination. In these cases it is best to administer the anti-serum, without wating for the meningococcus to be found. I am confident that I once saved the life of a physician's child by acting on this principle in a case of cerebrospinal meningitis, although I thought when I saw the fluid at the time of the first lumbar puncture that it was tuberculous meningitis. It proved to be an isolated case of meningococcic meningitis of severe grade, yet the fluid was clear throughout and leukocytosis was not present, and only after long search were one or two rather doubtful diplococci found. Nevertheless the case reacted favorably to every injection of the serum and eventually made a complete recovery without sequelæ.

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Hemorrhage from the Spinal Meninges.—This is almost always of traumatic origin in childhood, although it may occur in hemophilia and in scorbutus. It may be epidural or subdural, or rarely intradural.

In children it is almost always due to falls on the back or to severe blows. It probably is not uncommon from obstetric trauma, but if so it does not cause symptoms that lead to its diagnosis. The only cases that I have seen with spinal symptoms, attributable to obstetric trauma, have presented a condition distinctly indicative of hemorrhage into the cord substance itself, and this is true also of traumatic spinal hemorrhage at any age, at least as far as clinical symptoms are concerned. Hemorrhage may also occur from new growths within the vertebral canal.

Epidural or intradural hemorrhage is more likely to produce localizing symptoms than hemorrhage into the subdural space. The symptoms are those of local nerve-root irritation and if the hemorrhage is severe or progressive, eventual symptoms of compression of the cord supervene, and a localized meningomyelitis may result. Compression of the cord rarely goes so far as to produce lower motor segment symptoms at the site of the hemorrhage, but upper motor segment phenomena below the level of the hemorrhage, in the shape of varying degrees of spastic paraplegia with heightened reflexes and the Babinski phenomenon, and possibly ankle clonus, may be present, due to pressure on the pyramidal tracts at the site of the lesion. More or less spinal rigidity may exist. Pain and paresthesic phenomena corresponding to the nerve-roots at the level of the hemorrhage may be complained of by older children. Some degree of impairment of sensation below the level of the hemorrhage may also exist, but any considerable degree of implication of the sensory conduction tracts of the cord should always excite suspicion of hemorrhage within the substance of the cord itself.

Sphincteric disturbances, usually of the spastic type—retention of urine, difficulty in starting urination, dribbling of urine and constipation—may occur.

The onset of these symptoms may be abrupt but are usually gradual, depending upon the volume of the hemorrhage and the degree to which it is localized.

Subdural extravasation of blood may produce similar phenomena, but this variety of meningeal hemorrhage is more apt to produce general phenomena of meningeal irritation which are indistinguishable from the phenomena of meningitis, which in fact usually appear eventually.

Prognosis.—The outcome depends chiefly on the cause of the hemorrhage. Hemorrhage from the meninges in the cervical region however, regardless of its origin, is frequently rapidly fatal but the greater number of such instances in childhood are secondary to severe trauma with injury of the cord itself. This will be referred to later. Under any circumstances however the prognosis is necessarily grave and death may occur early or it may result from secondary meningitis later on.

Diagnosis.—In the event of known trauma the appearance of irritative spinal phenomena may well give rise to suspicion of hemorrhage, but a definite diagnosis cannot be made for these phenomena may be due to fracture, dislocation, or even the effects of spinal concussion, with secondary edema. When the hemorrhage is subdural lumbar puncture will of course reveal the presence of blood in large amount, but this will not exclude the possibility of coincident injury to the cord or to the nerve-roots. If hemorrhage alone exists the symptoms, as a rule, are gradual in appearance and progressive, whereas in a crush of the cord the symptoms are immediate, and usually complete at and below the level of the lesion. Even here, however additional symptoms may supervene due to advancing edema of the cord or to extension of a hemorrhage within its substance. At the best therefore the diagnosis of spinal meningeal hemorrhage is extremely difficult.

Treatment.—If epidural or even subdural hemorrhage can be definitely determined, operation may be considered, especially when its occurrence is secondary to trauma. The condition of the patient is often such however as to render the operation of laminectomy extremely hazardous. This will be referred to later in the consideration of the treatment of hematomyelia.

Ice-bags to the spine, venesection, local cupping are of very doubtful value. The same may be said of remedies to increase the coagulability of the blood but the latter may be tried if the symptoms are slowly progressive and seem to indicate a slow hemorrhage.

TUMORS OF THE SPINAL MENINGES AND OF THE SPINAL CORD

Tumors involving the functions of the cord may be situated in the vertebræ, in the meninges or in the cord itself. All forms are exceedingly rare in childhood.

Among the new growths in the vertebræ that have been reported as producing cord symptoms in childhood are sarcoma, fibrosarcoma, fibroma, fibromyoma and endothelioma. It is also conceivable that multiple exostoses, several instances of which have been reported in children might also produce spinal symptoms. Tumors of the spinal roots themselves, multiple neuromata, may occur in the cauda and cause radiating root pains and possibly the flaccid type of paralysis in the legs.

The tumors of the bone produce the cord symptoms usually by pressure or by root-irritation, but may invade the cord itself.

The phenomena produced by root irritation and by pressure are the same that occur in tumors of the meninges.

The most common tumors of the meninges in childhood are tuberculomata and sarcomata. They may be extradural or intradural or both. Several instances are on record in which sarcomata, apparently starting in the dura or external to it, have not only penetrated the dura and have involved the leptomeninges, but have also penetrated the interveterbral spaces and have formed tumor-masses evident on external examination, without involving the vertebræ.

Sarcoma is usually secondary to sarcomatous growth elsewhere. It may however be primary. It sometimes takes the form of diffuse infiltration of the meninges, extending more or less throughout the length of the spinal canal. Sommerfelt has recently reported an instance of sarcoma originating in a spinal ganglion with secondary infiltration of the pia, cord and brain, and presenting the clinical picture of ascending paralysis.

These are instances of sarcomatosis. They are usually metastatic.

Fibrosarcoma and gliosarcoma have also been reported in childhood, the latter form metastatic from the orbit.

Sarcoma is chiefly seen in the later years of childhood.

Tuberculoma is invariably secondary to a tuberculous focus elsewhere than in the nervous system, usually in the lymph glands or in the lungs. It seems extremely unusual as a result of tuberculosis of the vertebræ, miliary dissemination in the meninges with local or general tuberculous meningitis being the usual meningeal implication in such cases.

The tuberculoma is always formed of an aggregation of tubercles which coalesce and ultimately become indistinguishable as individual tubercles. This conglomerate tubercle forms a well defined tumor-mass, often encapsulated, and usually solitary. They may be multiple in the spinal canal or they may be associated with similar intracranial tumors, especially of the cerebellum.

They are more likely to occur in the first decade of childhood, and wherever symptoms of cord pressure occur in children, in this period of life, with a discoverable tuberculous focus elsewhere, Pott's disease or spinal tuberculoma should be thought of.

These tumors are somewhat more common at autopsy than clinical statistics would indicate.

Other tumors of the spinal meninges are too rare to require special mention. Circumscribed gumma of the meninges is unknown in childhood. Epidural or subdural lipoma is occasionally met with but more often found post mortem than discovered clinically. It probably exists without producing cord symptoms. Several instances of lipomatous tumor-masses have been reported in connection with spina bifida, sometimes protruding for a considerable distance.

These lipomata are benign, and originate in the epidural or subdural fat.

Endothelioma is very rare, but a case is on record in a 12 year old boy, involving the vertebræ and the dura.

Of tumors in the cord substance in childhood, tuberculoma, sarcoma and glioma are in frequency in the order named.

Tuberculoma forms usually a well defined and often encapsulated tumor-mass. About the mass the cord substance usually shows more or less softening and usually a zone of inflammatory reaction. The remarks in connection with similar tumors of the meninges apply to tuberculomata of the cord itself. The invasion is apparently through the arteries, especially through the branches of the anterior spinal artery, and thence the growth of the tubercles is outward toward the circumference of the cord, until by aggregation and fusion a conglomerate tumor-mass is formed. This, like all conglomerate tubercles, shows a tendency to central caseation and softening, although the softening does not always occur. Like tuberculoma in the meninges this invasion of the spinal medullary substance is chiefly seen in early childhood.

Sarcoma of the cord usually forms a well-defined tumor-mass, of softer consistency usually than the tuberculoma. It is also more apt to extend for a greater distance longitudinally.

What has been said in regard to sarcoma of the meninges applies also to sarcoma of the cord. Histologically it may appear in any of its forms. Its growth is usually rapid.

Gliomata usually originate in the ependymal and peripendymal tissue about the central canal of the cord, thence extending towards the periphery of the cord. Its growth is very slow.

While often extensive longitudinally through the cord, its transverse extent is greater at certain levels of the cord than at others. It is at these levels of concentration of the gliosis that transverse destruction of the cord structure occurs.

The newly proliferated tissue is often extremely rich in blood-vessels with very thin walls and consequently hemorrhages may occur, producing sudden signs of a transverse lesion, simulating the picture seen in acute transverse myelitis.

The gliomatous tissue varies considerably in histologic character and in rate of growth. In some cases it is rich in glia cells, in others poorer in these elements. In some instances the growth is extremely slow and extensive cellular degeneration may result in cavity formation with the pathologic picture of syringomyelia. The consideration of this form of gliosis does not belong here.

Symptomatology.—Tumors of the meninges and cord are sometimes found at autopsy where not suspected during life. Even where clinical signs eventually appear there has usually been a considerable period of new growth before distinct interference with cord function takes place.

In tumors originating in the vertebræ and in those of the meninges the earliest symptoms are usually those of posterior root irritation, especially pain, following the peripheral distribution of the nerve fibers that pass through the particular roots affected.

This pain is usually of neuralgic character and often severe, but in children apparently much less so than in adults, and not infrequently it is a source of little or no complaint.

The pain is usually paroxysmal and irregularly intermittent, varying periods of freedom intervening between the attacks.

In the beginning it may be unilateral. These early pains are of great importance in the localization of the tumor. It must be admitted however that these typical radicular pains are not always present even in the adult and are quite often entirely lacking in the child. Their absence therefore is of little negative value. Pain is not often complained of over the site of the tumor. Paresthesia is not complained of. It is in fact rare in adults. Irritation of the anterior motor roots by intervertebral tumors has not been reported in childhood. It also is very rare in adults.

When it occurs in the adult it takes the form either of localized muscular twitching, or of more intense cramp-like contraction of entire muscles or muscle-groups.

Later in the development of the tumor paralytic phenomena due to abolition of spinal-root function appear. The phenomena consist of the various forms of loss of sensory perception or of the lower segment type of motor paralysis, or both.

In the case of the abolition of sensory perception the upper level of the anesthesia indicates the highest level of the tumor. In the case of flaccid paralysis the muscles involved correspond to the site of greatest root involvement and consequently indicate accurately the location of the tumor, or at least the site of its maximum intensity. This is at least true of tumors in or adjacent to the meninges, whether extra or intradural, and it is these tumors that produce root symptoms, and these symptoms appear comparatively early in the tumor's development. Usually however before the appearance of the sensory or motor paralysis evidence of interference with conduction through the pyramidal tracts, where they pass the site of the tumor, occurs. This consists of the gradual development of spastic phenomena in the legs, never in the arms, although it is conceivable that this might occur in a tumor situated very high in the cervical cord.

This spastic paraplegia of course does not occur in tumors of the conus or of the cauda. This spasticity is due, as a rule, to direct pressure on the cord, but in the case of intra-medullary tumors it may occasionally be due to implication and partial destruction of the pyramidal tracts. The first sign of this condition is an exaggeration of the tendon reflexes. Quite often this will be found soon after the onset of the root pains. Such a combination of root pain above with increase of deep reflex activity below should always suggest spinal tumor.

In tumors having their origin within the cord itself root symptoms are not present unless the growth is located in the root zones, in which case irritative or paralytic phenomena may occur as in extra-medullary tumors.

Tumors involving the cauda produce nothing but root symptoms, and pain is a conspicuous feature of the case. The motor paralysis if it occurs is of course always of the flaccid type.

It is obvious that the particular combination of sensori-motor paralysis presented by each individual case of spinal tumor depends on the situation of the tumor and cannot be presented in any general consideration of the symptomatology. It would in fact require an entire reconsideration of the subject of spinal localization.

The essential facts are the signs that suggest spinal tumor. Its localization depends upon the following out of the principles described in the section on spinal topography and segment localization.

The most common site of spinal tumor in childhood, when it does not originate in the vertebræ, is the lower cervical region.

Pain or lower segment paralysis in the arms, often unilateral at first, are the initial symptoms in such cases, and later on spastic symptoms in the legs appear, these also at first sometimes unilateral, on the same side as the arm first affected.

It is well to keep in mind that the clinical symptoms do not necessarily indicate the actual extent of the tumor. This is especially so in gliomatosis or sarcomatosis of the cord.

Diagnosis.—The appearance of radiating pains limited to the territory supplied by one or two of the spinal roots, soon accompanied by heightened reflex activity in the limbs below, should always suggest spinal tumor if there is no evidence of disease of the vertebræ. Vertebral tuberculosis often makes its presence first known by these signs and it is far more likely to be their cause than tumor. In the great majority of such cases by the time these symptoms have appeared some degree of deformity will be noticeable in the vertebral column, or if this is not present, pain and rigidity will be elicited by attempts to passively move the spine, while a radigraph will finally clear away any remaining doubt as to the condition present.

If the early root pains of spinal tumor are absent the diagnosis may be impossible for a considerable period. Sooner or later however diminishing sensory perception appears, and the upper level of this hypesthesia is fixed and does not, as a rule ascend to higher segments. This lessened sensory perception corresponds at first to one or two spinal roots, sometimes unilaterally, but eventually on both sides, and soon increases to total sensory loss at that level, with abolition of all reflexes whose sensory arcs pass through those roots.

By this time diminishing sensation in all of the body below this level will usually be apparent, preceded, as mentioned above, by increase of the deep reflexes and spastic signs in the legs. Either at this time or sometimes much earlier, even preceding the sensory phenomena, flaccid paralysis may appear. It usually corresponds, in its location and distribution, to that part of the transverse area of the cord at which the destruction has been most extensive. In childhood this is likely to be in the upper extremities, owing to the frequency of involvement of the cervical cord.

There is no other condition that presents quite this slowly developing clinical picture. The nearest similarity in the adult is produced by syphilitic meningo-myelitis. This is rare in childhood. The only immediate method of differentiation consists in lumbar puncture and examination of the spinal fluid for the Wassermann reaction and for lymphocytosis.

Very rarely the first symptoms of tumor appear suddenly, either through hemorrhage into a previously latent tumor, or through sudden edema around the new growth, or on account of sudden increase in its rate of growth. It may be impossible, at first, to differentiate the condition from acute transverse myelitis, which would naturally be the provisional diagnosis under such circumstances. After the first few weeks however the objective symptoms of myelitis begin to show improvement which is progressive up to a certain point. In tumor of the cord, on the other hand, the objective signs show steady progression, although it may be slow. Finally, it should be remembered that transverse myelitis shows a predilection for the dorso-lumbar cord, whereas the cervical cord is the most common site for tumors in early life.

Prognosis.—The outlook is hopeless unless the tumor is extramedullary and easily accessible to removal. Naturally if at the same time it also happens to be extradural the chance of successful intervention is immeasurably increased.

Treatment.—The sole treatment is removal of the tumor. Whether this is feasible or not can be determined only by laminectomy and direct inspection of the character and location of the tumor. Every operation in spinal tumor is therefore of necessity an exploratory procedure.

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HEMATOMYELIA

Hemorrhage into the spinal cord in childhood is almost always due to trauma. Cases have been reported in hemophilia, in purpura hemorrhagica and in pertussis but these instances are extremely rare.

Obstetric injury of the cord is rare. It is of course not always recognized, or the condition is attributed to other causes. Spina bifida "occulta" and "intra-uterine poliomyelitis" have much to answer for in concealing the truth in this connection.

Postnatal trauma of the cord is extremely common in childhood as in adult life. In practically all of the cases of fracture of the spine, where cord symptoms are present, hemorrhage into the cord has occurred. Even more common perhaps are the cases where no fracture of the vertebræ is present nevertheless severe lesion of the spinal medulla is very evident. These cases are due either to a momentary dislocation of vertebræ at the time of the accident with crushing of the cord, or they are due to hemorrhage into the medullary substance, often to both factors.

Symptomatology.—Hemorrhage due to severe spinal trauma often presents the symptoms of a transverse lesion of the cord, not perhaps from the hemorrhage alone but from crushing or bending of the cord at the time of the injury. There may or may not be a fracture or displacement of the vertebræ. I think that in many of these cases there has been displacement of only momentary character with consequent crushing or bending of the cord. At any rate it often happens that by the time the consultant sees the case there is merely the evidence of a cross lesion of the cord and no evidence of vertebral abnormality. It has been my fortune to see many of these cases on the consulting service of a large general hospital and it is usually possible to account for the lesion only on the basis of an extensive medullary hemorrhage. I have no doubt however that there is not infrequently an actual crushing of the cord. These cases show a practically complete loss of function, sensory and motor, below a certain level, usually lower cervical or lower dorsal.

In less severe instances it is possible to determine a more limited lesion, more or less restricted to the gray or to the white matter of the cord. It is particularly within the gray matter that hemorrhage is likely to occur. It is then usually bilateral and extends for some distance longitudinally through the cord, but often unequally so.

The sensory and motor paralysis is then somewhat asymmetrically distributed. In these hemorrhages within the gray matter, the motor paralysis when it occurs is of course always flaccid. There may be dissociation of sensory disturbance, certain forms of sensation being lost while others are intact. It is particularly pain and temperature sensation that are implicated.

Rather rarely hemorrhage may chiefly or exclusively be located in the pyramidal tracts. Spastic symptoms then appear in the parts below the lesion. This may be unilateral.

Multiple hemorrhage may occur and under such circumstances there may sometimes appear a flaccid paralysis in one locality with spastic signs elsewhere.

All possible combination of cord symptoms may occur, depending on the location and extent of the hemorrhage.

Brown-Séquard paralysis may occur, due to unilateral hemorrhage.

Diagnosis.—In the absence of trauma, hemorrhage into the cord is a difficult condition to diagnose. The sudden onset of severe cord symptoms during the course of affections, known to be characterized by a tendency to hemorrhage in general, affords the basis of at least a presumptive diagnosis.

Hemorrhage without trauma and without apparent etiologic factors cannot be diagnosed clinically. Such cases would naturally be attributed to myelitis, and there is complete similarity in the cord symptoms. Rare as is transverse myelitis in childhood, hematomyelia without preceding trauma is even rarer, so that difficulty in diagnosis is not often likely to occur.

In traumatic cases the question often arises as to whether the symptoms are to be attributed to a crush of the cord or to hemorrhage within its substance, or to both conditions. This question often cannot be answered. Even if examination fails to reveal either fracture or dislocation, a crush or torsion of the cord may nevertheless have occurred. In most of these cases hemorrhage is probably a contributing factor.

While many instances of hemorrhage produce the clinical picture of a cross lesion of the cord, similar to that produced by a crush, any marked asymmetry in the symptoms suggests hemorrhage rather than a crush of the cord. Lighter degrees of hemorrhage accompanying trauma are not likely to offer any difficulty in diagnosis. I have seen the diagnosis of anterior poliomyelitis made in cases where the symptoms appeared immediately after a fall. It is quite possible that some of these instances were due to circumscribed areas of hemorrhage. Such cases are free from the general febrile phenomena of anterior poliomyelitis, although of course the absence of these phenomena is common enough in the latter affection.

Pathology.—The hemorrhage is usually into the gray matter. It may be in multiple foci after spinal concussion, or it may be in but one locality. It is apt to extend through several segments of the cord, especially in the gray matter, and may exceptionally work its way through the cord longitudinally for a much longer distance. Very rarely does it break its way horizontally through the cord to the meninges, except in a severe crush of the cord. About the hemorrhagic focus softening usually occurs, due to a secondary myelitis. The extravasated blood may be absorbed leaving a cavity and sometimes cyst formation or there is simply a proliferation of neuroglia taking the place of the destroyed medullary substance.

Prognosis.—The immediate prognosis depends upon the severity of the causative trauma and upon the site of the hemorrhage. If the latter is in the cervical cord the immediate outlook is less favorable than when it is in the dorso-lumbar cord. Secondary softening may extend upward to the phrenic centers and respiratory failure closes the scene. This is almost invariably the case in those instances where the clinical picture immediately after the trauma indicates a complete transverse cutting off of cord function from the fifth or sixth cervical segment downward. These cases are found with entire abolition of thoracic respiration and breathing only with the diaphragm. In the course of a few hours this also ceases, sometimes quite suddenly, and terminates the case. In lower lesions of similar severity, there is usually left a total paralysis below the level involved, with the subsequent development of cystitis, pyelo-nephritis and bedsores and eventual death from sepsis or bronchopneumonia, etc.

When the symptoms are not so severe and the signs indicate an incomplete cutting off of the conduction tracts, the outlook is much more favorable and life is not necessarily endangered. These cases however are left with more or less permanent disability.

Treatment.—The securance of absolute immobility, even in the lightest cases, if feasible in the prone position, although the latter is often impossible, is the most important indication. If the prone position is not contra-indicated, advantage may be taken of the position to apply ice-bags over the spine. Owing to the chief source of the arterial supply to the cord and the depth at which the vessels lie below the surface of the body it is extremely unlikely that this attempt to influence the flow of blood is of the slightest effect. Usually in traumatic cases the maintainance of this position is impossible. The peculiar conditions of spinal arterial circulation also make it unlikely that any internal medication with the idea of reducing blood pressure sufficiently to influence the hemorrhage will avail. Moreover the worst has usually happened by the time the case comes under observation, and the future depends upon the degree of destruction, or on the occurrence or non-occurrence of secondary edema or softening, neither of which can be influenced in any way by treatment.

Animal experimentation has shown it to be feasible to relieve the effects of traumatic hemorrhage by slitting up the cord longitudinally in the region affected.

The treatment of the paralysis and secondary contractures of the later period of such cases as do not succumb differs in no way from that employed in similar conditions of different etiology.

CHAPTER CLXXXV

THE PSYCHOPATHOLOGY OF CHILDHOOD

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INTRODUCTION

A clear-cut and relatively well-organized statement of psychopathologic issues such as is possible to a very large extent in connection with adults is still an unrealized ideal in the domain of childhood. But this state of affairs has its advantages as well as the handicap of rendering exceptionally difficult a ready orientation in the field of childhood psychopathology, for in many respects it would be unfortunate to follow uncritically in dealing with these disorders of childhood the traditional tendencies, concepts and elaborate schemes of classification of adult psychopathology. Much of this has come to be recognized as a serious stumbling block to progress and much of the energy in the field of psychiatry is being at present devoted to a re-examination and revaluation of its traditional hypotheses of causation, outcome and management. In the main, the present-day ideal, as particularly championed by Meyer, is in the direction of a simplification of issues and of a greater emphasis of the rôle which ordinary, every-day events of life may play in the adaptive difficulties of man. Moreover, while a thorough grounding in psychologic and pathologic theory and an acquaintance with the various philosophic notions of life is recognized as a very valuable asset in dealing with the problems of psychopathology, the management of the concrete case demands above all else a sound, common-sense attitude toward life as it is actually reflected in the common, every day pursuits of man.

The soundness of these newer tendencies receives special corroboration from the field of Childhood-Psychopathology. Here the issues met with, because they are less often in the nature of "end-results" of a long process of disintegration, either physical or mental, impress one particularly with their simplicity and the relative naturalness of the disorder as a "reactive manifestation" on the part of the individual to certain events or situations in his life-experience as a biologic and social unit. The necessity in this connection of viewing the child as an integrated unit is paramount, and one of the important by-products of the reaction from the traditional psychiatric concepts has been the renewed effort to counteract the unhealthy tendency of modern medicine to view the problems of human disease and disorder in what might be termed a "fractional" or "part-function" manner. The high degree of specialization which characterizes modern medicine carries with it as an inevitable by-product an altogether too frequent neglect of the patient as a personality, as a biologic and social unit, with pursuits, problems and frustrations of the "man as a whole," aside from those which might be the primary or exlcusive concern of the specialist in stomach or heart diseases. It is only exceptionally that the surgeon or internist seeks to discern in the problems of the "personality as a whole" an explanation for the specific disorder of an organ or system of organs which he is called upon to diagnose and treat, and in the curricula of many medical schools very little time and effort is devoted to an understanding of the human being as an integrated whole, charged with the tasks of meeting adequately not only the needs of his organic and biologic nature but also those adjustive requirements of a psychologic and social character which life in contact with others constantly brings with it.

Thus the patient's surgical or medical complaint may be merely a way of expressing a much deeper and more inclusive problem of the personality, or it may be merely a tool, an instrument by means of which the patient hopes to achieve a more acceptable adjustment to his problems of biologic or social adaptation.

A full recognition of the relative inadequacy of what have been termed the "impersonal categories" of surgery and internal medicine is an indispensable prerequisite for a proper orientation in the subject to be discussed in this chapter. Whether one is dealing with the mere tantrums of childhood, or with the various substitutive phenomena in the nature of vomiting, headache, convulsive manifestations, etc., or with the more pronounced anxiety states and obsessional thoughts and acts occasionally met with in children, or with the more socially significant delinquency manifestations of stealing, lying or running away, it is essential above all to search for the meaning of the disorder in the child's economy, for the rôle it plays in the child's efforts to adapt himself to the demands of his environment. This view of the matter does not involve in the least a neglect to recognize the many childhood manifestations of a neurotic or psycho-pathologic nature which may have their roots in more or less definite constitutional disorders of an inherited or acquired origin, or of the rôle which physical or toxic agents play in the psychopathology of childhood. Some of these strictly defined nervous and mental disorders of childhood which can be more definitely understood on the basis of a strict "medical pathology" have already been dealt with elsewhere in these volumes, while others will be considered in this chapter. The point that needs emphasis is that even where one is dealing with issues which can be explained in terms of an underlying constitutional or more accidental pathologic factor, the element of personality and of the situations of life surrounding the individual plays an important rôle and exerts a modifying influence on the manifestation of the disorder as well as on the results of therapeutic endeavor. These considerations, which apply to the adult likewise, assume a greater significance in children for reasons which are closely related to the very nature of the childhood state of dependency and relative impotence. More so than is apt to be the case with the adult, the child's striving for recognition and for an enlargement of his personality renders him subject to a utilization of the element of disease in the service of the ego. Equally purposive may be the various manifestations of disordered behavior, a clear understanding of which can only be had when they are viewed as attempts on the part of the child at some sort of adjustment. That these attempts at adjustment may actually lead to serious maladjustment only strengthens the argument for the need of a thorough understanding of the rôle which the child's reaction plays in his personal economy.

Frequently, what the adult considers as an unhealthy or perverse manifestation of childhood behavior may be nothing less than the child's reaction to an imperative instinctive demand which must gain expression somehow, and it becomes our task not to thwart this energy expression by blind restriction but to guide it into healthy channels of activity. Moreover, the student of psychopathology must always consider the possibility of an unconscious element in the reaction of the personality as a whole, so that the overt behavior manifestation may only remotely and symbolically reflect the deeper, underlying situation to which it is related. The failure, therefore, to recognize a definite meaning or purpose in a symptom or a behavior tendency from a cursory examination of the situation does not always justify attributing the difficulty to some obscure constitutional or hereditary factor. It is precisely this tendency to relegate the various behavior manifestations of the child or adult for which no ready explanation can be had to some innate and original fault or perversity of makeup that has retarded progress in this field. A related and equally stultifying tendency is reflected in the craving for cataloging and labeling which has unfortunately become identified with progressivism in medicine.

Aside from the difficulties frequently encountered in discovering the true meaning of a behavior manifestation, one commonly meets with a multiplicity of causes back of a psychopathologic disorder, in which hereditary and constitutional elements may play a rôle, but in which numerous events in the life of the individual may be found to have been operative in causing the condition. Actual experience leads to an increasing emphasis being put upon these "conditioning influences" of life-experiences and to a lessened willingness to be guided in the management of the problems of childhood psychopathology by theories of heredity and predetermination. This changing attitude stimulates a more spirited endeavor to understand what is going on "behind the scenes," so to speak, and a more experimental attitude as regards therapeutic endeavors. Specifically, in dealing with these disorders of childhood it is frequently, indeed quite commonly, necessary to deal with situations in which all sorts of factors and all sorts of persons may be involved besides the child. Of these situations and persons, the home and the school, the parents, brothers, sisters, teachers, and playmates assume a particularly important rôle. The child's intimate contact with a nervous, harrassed and over-burdened mother, or with an irritable, dissatisfied and moody teacher, or with an over-aggressive, overly successful brother or sister may condition types of reaction of a faulty or pathologic kind which become as deeply rooted and as determining for the personality development and behavior of the child as anything which it may have inherited, and a vicious circle may thus be started which is with difficulty only differentiated from some self-perpetuating hereditary factor.

A situation not uncommonly met with in the conjugal maladjustment of adults is quite illustrative of this. The mother, who because of a frustrated conjugal life of her own, turns to her son as a source of substitutive emotional satisfaction and thus cultivates in him an unduly exaggerated filial attachment which he carries into his adulthood, is apt in turn to render him incapable of a healthy, normal and satisfying conjugal career. Thus, he becomes conditioned to seek substitutive outlets and is apt to find them in a cultivation of an undue attachment to his daughter, if he should have one, and to continue the vicious circle into the third generation. There is no question here of heredity in the accepted sense of the term, but superficially the various neurotic manifestations which go with a frustrated conjugal life, reflected as they are in parent, child and perhaps grandchild alike, may readily be mistaken for an evidence of an hereditary neurotic taint. Indeed, much, if not most of what we meet with in the psychopathology of childhood, is quite explicable on the ground of unhealthy conditioning influences of life and the cultivation of unhygienic habits of biologic and social adaptation. This is one of the reasons why any attempt at a complete and inclusive classification of these manifestations of childhood is apt to be unreal and not at all expressive of the actual situation.

Another reason lies in the fact that in dealing with children well-delineated disease entities such as one is familiar with in adults are met with only exceptionally. Commonly, both in free dispensary work as well as in private practice, the problems one is called upon to solve are those of rather ordinary difficulties of adaptation to the demands of daily life, maladjustments which, to be sure, may prove to be very refractory to treatment and may cause parents and teachers much anxiety, but which are nevertheless quite understandable in terms of ordinary "household troubles," requiring none of the traditional complexity of clinico-pathologic terminology for a statement of the issue. To illustrate: Little A, barely seven years of age, is brought to the clinic by a distressed and excited mother who complains that she has reached the end of her endurance with the boy. He has been growing steadily worse in his obstinacy and in his "apparent love for doing just the opposite of what is wanted of him." Pleading with him is of no avail, in fact seems to exaggerate the tendency, and punishment, which according to the mother's admission has at times been rather severe, is equally useless. The boy will persist in teasing his younger sister, and seems to take delight in thus upsetting the peace of the household. He will continue to stay out late at night enjoying apparently the anxiety which it causes the parents, is obdurate in his refusal to keep himself clean and to be in any degree regular in the taking of food. When called to task or reprimanded he begins to howl at the top of his voice, until the neighbors become annoyed and excited, when he has to be bribed in some way into silence. Every scene practically ends in this fashion, with the additional conviction on the boy's part that he can be master of the situation and that his particular "technique" for being the center of attraction not only of his own household, but of the neighborhood as well, is quite an adequate one. Now the curious thing about the entire situation is that this obstinate and unmanageable boy at home is quite a model pupil at school, exceptionally well endowed in certain respects, and easily manageable in his conduct. Apparently, the thing that he is endeavoring to achieve at home, namely recognition, by means of obstinacy and terrorism he gains quite readily at school through his exceptionally good scholarship in certain fields. But unfortunately the home does not recognize these values, while the childish cuteness and gentleness of his litte sister is quite enough to absorb the entire attention of this emotionally starved household. After the mother was induced to take a little interest in the school activities of her boy, to talk over with him daily the experiences of his school life, in short, to give the boy an opportunity to "put himself across" with her in ways in which he was certain of success, the entire trouble at home practically disappeared and the mother discovered a new personality in her wayward boy.

Or take the case of a little girl of 12 who was brought to the office by a distraught and panicky mother because the child had been manifesting various signs of anxiety and a tendency to self-accusations of a vague and general kind. She would frequently get up at night and look under the bed and in corners of the room in an apprehensive manner, would have to be assured by her mother that she had behaved well that day, that she had committed no serious breach of etiquette or sin, etc. She was also doing very poorly at school and was threatened with dismissal from a private school which she attended because of her poor scholarship.

The child belonged to a well-to-do family of excellent social position and of a high order of intelligence, had received all the care and attention in her bringing up which money could secure, but had missed in her development the type of intimate contact with someone in her household in whom she felt she could implicitly trust and to whom she could freely unburden herself.

When this condition was provided in the consultation room the child told a story of how two years previously, when she was barely 10, two girl companions of the same age and a boy a little older had aroused in her a good deal of sex curiosity and introduced her to practices which mystified and bewildered her because of the strange sensations they gave rise to. In connection with these practices which she had been continuing from time to time since then, she developed a deep sense of guilt and anxiety, losing frequently considerable sleep at night and very often being pre-occupied with these thoughts during the day and while at school. It was this pre-occupation with a troublesome and distressing mental content which was responsible for her inattentiveness and consequent failure at school and not lack of intelligence. With the opportunity to talk these matters out freely in a sympathetic setting, and with the acquisition of such enlightenment on the subject of sex as was indicated, the girl became freed entirely of her anxiety state and showed considerable improvement in her school performance.

Now these brief reports of two rather dissimilar situations are sufficiently detailed to indicate that in each instance we were dealing with what was actually a very serious problem of maladjustment and where continued misunderstanding of the situation might have led, as is the case, unfortunately altogether too frequently, to a more serious aggravation of the problem through the application of wrong therapy. In the case of the boy, the mother had been frequently on the verge of calling in the aid of the police, which might have resulted, as it frequently does, in a type of contact with and misconception of "authority" which is found to be the starting point of many a delinquent career. In the case of the girl, on the other hand, it is difficult to overestimate the deleterious effect of the "sense of failure" which the dismissal from school might have engendered in her, particularly because of the sensitiveness of her family setting to such issues. And yet, serious as these problems were, they contained nothing which is embraced in any strict definition of "medical pathology" in the traditional sense of this term. Neither can these cases by any manner of means be forced into the familiar categories of current "pyschopathologic systems." One deals here with perfectly common and everyday childhood difficulties which require for a correct elucidation and management rather a better understanding of the "business of living," of childhood economy, both internal and external, than is furnished by current treatises on medical pathology. In other words, one would be closer to the truth in speaking of these problems as problems in psychobiology rather than in "medical pathology" in the traditional sense of this phrase.

Much more promise for a helpful orientation lies in an endeavor to understand the nature of the "child's personality," in so far as child-life is expressive of this, and of the various factors which are operative in the molding of this personality and in determining its life reactions. It will be our aim, therefore, by way of an introduction to a discussion of the actual manifestations of childhood psychopathology to review briefly the generally accepted views concerning the native endowments of the human infant, to follow this with a more detailed consideration of the conditioning influences more commonly met with which either hamper or further the process of shaping and disciplining these native attributes of the child in the service of a healthy adulthood and to conclude this preliminary statement with a discussion of the outstanding mental functions involved in this process.

THE PERSONALITY OF THE CHILD AND THE SIGNIFICANT FACTORS IN CONDITIONING ITS DEVELOPMENT

Leaving out of consideration here the question of intellectual development, an aspect of childhood which belongs more properly to the domain of normal psychology, the childhood state, from the point of view of psychopathology, presents significant issues which are related more definitely to the instinctive-emotional aspects of life.

These issues have to do primarily with the child's state of dependency and with his task of adaptation to the problem of authority, with his necessity for a progressive recognition of the world of reality and the abandonment to the required degree of the strong childhood tendency to have recourse to an uncritical phantasy-formation, with the important problem of his affective relationships to his parents and other near relatives, and finally, with the task of a progressive integration of the personality with the more critical shaping of purposes and methods for their attainment. These problems of the childhood state are merely more specific elements of the general problem of growing up, in the solution of which there is an active play of factors which are innate in the personality of the child and of those which emanate from the child's environment.

The manner of adjustment to these issues is absolutely determinative of that aspect of the adult personality which we designate as "character," and in the maladjustments of the adult there is ordinarily reflected some fault in the early adaptation to one or more of the foregoing fundamental problems. The obstacles to the successful solution of these issues have received considerable illumination through modern psychopathology, and a consideration of them is indispensable for a proper understanding of the personality difficulties of childhood. At the same time it would be erroneous to assume a too exclusive psychopathologic point of view in this connection. Much of the credit for these newer teachings is due to men like McDougall and Shand, whose contributions in connection with the problem of instinct and its organization into the complex emotions and sentiments which enter to such a large extent in determining human conduct, have helped materially in rescuing the science of human behavior from the aridity of the academic psychologies.

Perhaps a greater debt still, psychopathology owes the psychoanalytic school, from which have come most illuminating contributions to the entire problem of human behavior in health and disease.

It is not necessary to enter here into a detailed discussion of the still much controverted and important question of instinct in man, but there is no reason whatever for deliberately avoiding this question as is done by some students of psychopathology. To do so would render impossible the proper understanding of the forces that determine the child's reactions to the problems of life and of the serious conflicts that have to be overcome in the growth from childhood to adulthood.

In practice, any inclusive estimate of the "individual as a whole," a procedure which has come to be considered indispensable in modern psychopathology, necessitates a reference to the subject of instinct, since to fail to do so leaves one without an adequate conception of the individual's attitude towards his most fundamental needs and strivings. The neurosis, or maladjustment of the adult or child, is never quite free from evidence of an involvement in the process of these fundamental dispositions, and to ignore them renders an adequate understanding of the patient impossible.

There are still significant differences of opinion among students of the subject concerning the precise number and nature of instincts in man, but it is thoroughly safe and practical to assume that they can be comprised under three headings, each determining a quite definite goal or set of goals in the life of the individual and each accompanied by specific feeling-tones which determine the individual's attitudes to life.

In the first place, there are those innate dispositions and strivings which are related to the organism as "self," in the broadest sense of this term; then there are those which relate to the organism as a biologic unit carrying within its organization the purpose to propagate itself and to insure the perpetuation of the race, and finally, man carries within him a set of dispositions which are related to his gregariousness and membership in the human family.

While the individual instinctive dispositions, so ably discussed by McDougall,[1] develop their capacity for expression and for influencing human behavior at different stages in the individual's development, the childhood state already gives evidence of the operation within the child's personality of those innate tendencies described above. How true this is of the sex components has already been discussed elsewhere in these volumes.[2] The laws which govern the operation of instinct may be discerned during childhood even more readily than it is possible to do in adulthood since the relatively incomplete integration of the childhood personality and the relative poverty of individual experience render it more difficult for the child to disguise the immediate effects of a balking of instinctive expression.

This is particularly well illustrated in connection with the child's urgent and insistent drive to "put himself across" somehow, to an enlargement of the childhood self and to the achievement of a satisfying sense of "security" in the face of his relative impotence and helplessness. In the maladjustments of childhood there is reflected very often a result of, or reaction to, a "balking" of these strivings, and symptoms acquire meaning and lend themselves to a more rational therapy when one discovers their relationship to these egoistic strivings of the child. The domination of egoistic elements in the child's personality undoubtedly has its justification in the child's need to overcome the helplessness of childhood, but it constitutes an important factor in his difficulties of adaptation to the problem of authority and in his attempts at satisfying his innate herd impulses.

These difficulties which go with the child's necessity to give full expression to his "egoistic" strivings as an indispensable part of the process of normal growth on the one hand, and to adjust himself to the growing demands made upon him by the environment for a restriction of the personality on the other hand, expose him to a type of fundamental maladjustment which underlies many of the problems of childhood, namely, a clinging to those adjustive mechanisms which served him so well during the infantile state of absolute dependency. The extreme attractiveness of this type of fixation at an earlier level of development, or of a regression to it as is manifested by certain adult neurotics, lies in the fact that in spite of its utter state of helplessness and dependency, life at this time is a complete indulgence, only less so in degree than the fetal state, and demands in return very little exertion of effort.

The absolute security and satisfaction which the mother's love and tenderness bring to the infant, the more or less absolute control of the environment as far as personal needs are concerned through the mere exhibition of signs of discomfort, render this state extremely attractive to the individual and he only begins to experience the unpleasant features of a state of dependency in connection with attempts on the part of the adults in his environment to "bring him up," to educate him. But growth, to which the child is impelled by ineradicable, innate dispositions, demands a progressive giving up of this comfortable and effortless state and the gradual acquisition of the ability to stand on one's own feet and to adjust one's desires to the facts of reality. Also, there is the necessity of cultivating the ability to postpone the gratification of desire in accordance with the demands of reality and of the ability to interpose a greater or less degree of deliberation between stimulus or impulse and reaction.

Now the innate dispositions most intimately related to the child's adjustment to his dependency-authority problem are the instincts of self-assertion and self-abasement, and the type of personality one ultimately develops depends very largely upon the kind of balance one has achieved between these instinctive dispositions.

To this adjustment is related the important question of "self-esteem" which enters so basically into the behavior of man, and that of the degree of one's dependence for one's comfort and efficiency upon the esteem of one's fellows. These issues are largely decided in childhood by the manner in which the child's problem of dependency is managed by the adults who surround him.

It deserves repeated reiteration that in the growth from infancy to adulthood there is nothing, or at most very little, that requires complete elimination. Adulthood is best understood as a more complete integration, as an enlargement of the childhood state and not as a complete displacement of it. In the growth from infancy to maturity there is present the same principle which may be discerned in the evolution of mankind, namely, that each stage of development contains the elements which make possible the next more complex and more complete stage. The management, therefore, of the childhood "egoism," even in its most asocial and amoral aspects does not involve a process of elimination but one of correction and adjustment, lest in our efforts to educate the child we rob him of some of the most essential requirements for a healthy personality development.

In managing this problem in connection with concrete issues of bad child-parent or child-teacher relationships, the psychopathologist is frequently confronted with the difficulty of having to answer the question, "Why should a parent or teacher deliberately wish to do harm to the child?"

The answer to this question is bound up with what is perhaps the most noteworthy and practical contribution of modern psychology. In the first place, it should be one's task to show the parent or teacher that behavior and attitude are not always, nor even frequently, results of deliberation. The motives back of a parent's or teacher's attitude or conduct towards a child may be entirely hidden from them, and may spring from sources identical with those which determine the child's conduct. It is not ordinarily the parent's or teacher's adulthood which makes difficult his management of the child but rather the infantile and childish elements in his own makeup that are responsible for this. Moreover, the more closely related the difficulties of managing a child are with attributes in the adult of which he is ashamed as an adult, the more obscured is his vision likely to be with reference to these attributes.

To argue, therefore, merely on the basis of objective issues, of which the parent or teacher is fully conscious, is ordinarily futile and unless he possesses a capacity and willingness to know himself he will ordinarily fail to understand the child he is attempting to guide or educate.

Whether one is dealing with the not uncommon difficulty of parents to give up the deep emotional satisfactions which they derive from keeping their offspring in a babyish and dependent state, or with some of the more ugly aspects of the exhibition of parental authority of a more positive nature, or with irritations in a love-starved, maladjusted teacher, etc., the problem for solution is primarily a problem in selfknowledge on the part of the adult. Frequently enough the adjustment of the child is successful to the degree to which a better understanding is achieved by the adult immediately concerned in the situation.

To return to the main trend of our discussion, in assisting the child in the management of his problem of dependency and in the adjustment of his egoistic strivings to the demands of his herd impulses it should be the aim of the adult to appreciate that both these tendencies which are apt to come in conflict in the child's endeavor to meet the requirements of life, have their roots in innate dispositions which must gain expression somehow, and that education actually means a "drawing out" of the child's potentialities and not their elimination or stultification. What is needed as part of the growing up process is the striking of a proper balance between opposing trends so that capacity for personal satisfaction goes more or less hand in hand with the progressive capacity for self-denial. The effects of a mismanagement of this important childhood problem will be discussed in greater detail later.

As regards the child's preference for phantasy-formation, the healthy regulation of which is an essential condition of growth, the same holds true. It is not a question of the total elimination of this tendency, since an important element of a successful adulthood is constructive imagination, a direct descendant of the imaginativeness of childhood. While fully recognizing the need for a progressive attention to the world of reality, a too sudden or too complete checking of the childhood recourse to phantasy is not only apt to interfere with adulthood but to render this type of indulgence unduly attractive.

One must keep in mind that for a considerable period after birth the individual is wholly governed by what has been termed the "pleasure-pain" principle of existence. During this time the individual's main interest in life is the avoidance of pain and the pursuit of pleasure. Reality is accepted only in so far as it is in accord with this pursuit. Whenever reality becomes painful, the imaginativeness of the child readily transforms a painful situation into a pleasurable one irrespective of the actual conditions of reality. How difficult the process must be of abandoning this "magic wand" of childhood is attested to by the readiness with which adults succumb to this adjustive mechanism. The gradual harnessing of this tendency to the facts of reality is not only a condition of healthy growth but is of importance in the child's immediate existence. The misinterpretations of environmental issues and relations so readily resorted to by children, misinterpretations which sometimes lead to the cultivation of thoroughly unhealthy attitudes to parents, sisters and brothers and to life in general, are closely linked with the child's great imaginativeness.

The ready identification of children with the heroes and martyrs of their environment is probably also dependent upon this imaginative faculty.

These issues assume even greater significance when taken together with the great imitativeness of children. It is not necessary to discuss here the rôle that imitation plays in the growing up process, and how much formal education owes to this imitative faculty of the child. But here again, as is the case with the necessity for the gradual substitution of a recognition of reality for mere phantasy formation, imitativeness which serves the child so well to acquire rapidly various elements in his social heritage must gradually give way to independence in thought and action.

Failure of a proper management of the child's imaginativeness, his imitativeness and related suggestibility leads to serious maladjustments both in the child and in adulthood. It might be added that these elements of child nature have an important bearing also on the manner in which the child solves his dependency-authority problem.

With reference to the affective relationships of the child to his parents and near relatives, much has already been covered in the chapter on the sexual life of the child. The rôle which the parents and near relatives, or substitutes for these, play in determining the child's affective life cannot be overstressed. More so even than is the case with the factors in child-life already discussed, the elements of conflict and obstacle to growth are here apt to be environmental in nature, and the required adjustment demands even more understanding and tact than is the case with the dependency-authority problem. The reason for this lies in the fact that the child's attitude toward the very important problem of mating, and his chances for a healthy sexual maturity are intimately bound up with his experiences in these early expressions of his love-life. Of the maladjustments of adulthood none presents such pitiable and almost ineradicable reminders of a mismanaged childhood as those which have to do with the child's love-life.

Maladjustments in this sphere are intimately bound up with the next important problem which we wish to touch upon, namely, the problem of personality integration. Now it is quite obvious that no element of child nature and experience is unrelated to this main problem of integration, since integration at any level in structure or time, is the process of putting together all the elements of the individual's structure and experience in the creation of the new whole. But the love-life of the individual constitutes the very core of the integrated whole, and colors to a very determining degree his main purposes in life. It is largely in accordance with the pattern of his childhood affective relationships that the individual shapes his love-life and his attitude towards the problem of mating. But this element of the child's affective relationships enters also into his immediate behavior as a child. The destructive emotions of hatred and jealousy relate in the child largely to the persons in his immediate environment rather than to the wider sphere outside the home, and are determined by the environmental responses to his exhibitions of love.

In thus briefly touching upon the several important elements of an instinctive-emotional nature which play a significant rôle in the behavior of the child, it is naturally not intended to convey the impression that any one of these elements can be understood or affected altogether singly, and without reference to the other aspects of child-life. Any attempt to view the child in such a fractional manner is apt to render understanding difficult, if not impossible.

For this reason, what has already been said should be supplemented by a consideration of the child's growth as an individual through infancy and childhood, a discussion which will be made possible in connection with the presentation of the common conditioning influences of child-life.

What has been said thus far with reference to the child's personality referred to elements which might be considered as common of all childhood. The problems touched upon are rather in the nature of potential factors which may lead to serious maladjustment under certain conditions. It remains for us to review the more commonly met with conditions which exert an influence for good or bad on the childhood personality and on the process of growth to maturity. There is some justification for beginning this presentation with the pre-natal state. Not that it is our intention to enter here into any detailed discussion of the subject of heredity. Outside of the more or less clear-cut issues in mental deficiency, or constitutional inferiority or frank mental disease, there is very little that can be said concerning the rôle of heredity in the maladjustments of childhood.

But with the emphasis already given the subject of environmental influence it must be evident that exposure of a child to life in intimate contact with an insane or profoundly neurotic parent may well exercise influences as deleterious to the childhood personality as anything that heredity might exert upon it. Moreover, one might emphasize more strongly than is ordinarily done, the evil of exposing the growing child to a knowledge that he carries an hereditary burden, especially where this is frequently reiterated and made part of a more general exhibition of anxiety concerning the outcome of a given child's career. A pre-occupation with thoughts concerning the possible expression in one's life of an hereditary burden may operate as a serious insult to the self-esteem of that person and may contribute to the development of a sense of inferiority or to the fixation of an already existing tendency in this direction.

It might be well to discuss more fully at this point the customary manner in which the personality reacts to insults which give rise to a feeling of inferiority. In the first place it must be made clear that in view of the helplessness and dependency of the childhood state coupled with the almost ineradicable human tendency to cultivate aspirations which are quite beyond the possibility of attainment, a certain degree of a relative inferiority feeling is probably characteristic of all mankind. Much of the energy of the ordinary human life is devoted to efforts at compensation of this sense of inferiority.

But in the case of individuals who have been exposed to a specifically serious insult to their self-esteem, the inferiority induced may lead to reactions which are of a determining influence upon behavior. If the insult is severe enough, especially when it concerns a particularly sensitive individual who is already more or less intimidated by the problems of life, the effect may be a crushing one, leaving the individual in a profoundly intimidated state and deprived of all capacity for any kind of aggressive attitude towards the problems of adaptation. Adults who suffer in this manner spend their days in an impotent state of anxiety and fear of the future, occasionally finding some solace in the cultivation of the bitter cynical attitude of the deeply disappointed man.

In children there may be in addition what might be termed a spread of the effect of the insult to their self-esteem beyond the immediate situation, engendering a functional inability or inferiority along lines in which they originally possessed good ability. The situation is well illustrated in those school children in whom intimidations outside of the classroom cause a deprivation of the necessary degree of self-confidence for good scholarship. The secondary discouragement of school failure only serves to accentuate the feeling of inferiority and humility.

We have already indicated earlier how absolutely essential for healthy adaptation is the cultivation of a satisfying sense of self-esteem. Insults to the personality are not always reacted to in the manner described above. In some individuals compensatory manifestations in the nature of an over-aggressiveness and wholly unwarranted self-confidence come to light which may be as hampering to the personality as are the manifestations of a pathologic degree of timidity.

The classroom bully and street hooligan are often expressions of a compensatory mechanism to a sense of inferiority. The matter stands even worse with those children who, developing neither of these types of reaction, resort to a compensatory day-dreaming in which life's problems are solved by merely imagining a solution. Occasionally the progressive ignoring of reality and the growing satisfaction which is derived from day-dreaming leads to actual delusional formation.

Aside from the inferiority feelings which may be induced by anyone of a variety of insults to the personality to be discussed later on, actual deprivations in the nature of physical defect or disfigurement may induce this feeling. It seems to be the case that where the inferiority feeling is based upon an actual deprivation of this sort it leads more often than not to compensatory efforts of a constructive nature, which may leave the individual in the end with an actually greater sense of self-esteem than that with which he has started.

The overhanging cloud of a knowledge that one harbors an hereditary burden with quite unknown possibilities for affecting one's future, especially so where it concerns neuropathic heredity, is quite capable of inducing a deep sense of inferiority for which a solution has been known to have been sought in suicide. It is of importance, therefore, in discussing the problem of heredity with tainted people to keep this possibility in mind. Certainly our limited knowledge concerning the hereditary transmissibility of neuropathic tendencies justifies at least an attitude of open mindedness, devoid of the tendency to make dire predictions so often exhibited by physicians who ignore the questions just referred to. Equally justifiable is a greater concern with the elements affecting the intra-uterine life of the individual. Particularly worthy of consideration is the emotional life of the pregnant mother and her attitude towards the expected child. The one, it has been definitely shown by physiologic research, has a definite bearing upon the nutrition and growth of the fetus, while the other might lead to the creation of a thoroughly unhealthy mental setting for the child which is not without serious consequences to his personality.

With the moment of birth significant changes of a psychologic nature take place in the personal economy of the infant, since from the moment of birth demands of one kind or another for adaptation to reality begin to assail him.

From the moment of birth, the new individual begins to be exposed to the countless influences which life brings with it and which go to determine the mass of memories, habits and resources which are employed in the task of adaptation.

The human infant, in spite of the wondrous potentiality which he carries within him to develop into the reasoning adult, is by far the most helpless and dependable of infants, and thus subject not only to his own laws of development but to experimentation with these laws on the part of untrained parents. The situation has become particularly aggravated in late years with respect to the "puttering" by untrained parents on account of the mass of trashy literature on the subject of bringing up children to which parents have access. Parents come to consult the physician with the lamentation that this or that pet formula for the bringing up of children which they have gained from some book on the subject has failed to work.

The important point that is usually ignored by these "Guides for Young Mothers," etc., is that no two children are quite alike in all respects, and no method of child rearing has as yet been devised which would fit all cases. If we perchance should ever hit upon such a universally applicable set of rules for the bringing up of children we would probably wind up, as someone has said, with a race of uniform dunces. The infant's first privilege ought rightly to be the opportunity to show what stuff he is made of, so that in a very significant sense it will be our duty to be wise and discreet in the matter of letting the newly born infant alone, give him a chance to show himself for what he is. This caution is necessary in view of the very active tendency on the part of those who have the guidance of the individual in hand, either by divine right or because of public regulation to "take hold and do something," and the earlier the better.

This, I need not say, cannot be interpreted even by the most experimentally minded as a "counsel of neglect." The positive thing that the parent can contribute at this time is to see to it that the child has a chance to become a stout little animal and to build up the resistances to withstand the dangers that lurk in his physical surroundings.

But there are certain psychologic elements even in connection with very early infancy which a wise parent will endeavor to keep in mind. We have referred already to the infant as a pleasure-seeking and pain-avoiding animal which ignores reality. Part of this reality which the imperious infant refuses to recognize is the comfort and peace of mind of his parents. It may be difficult to train a child to take nourishment or to attend to calls of nature at certain convenient intervals, or to go to sleep naturally without the necessity of the lulling influence of some monotonous stimulus, like the swinging bed or cradle, or the thoroughly vicious habit which some parents resort to in putting their babies to sleep of stroking or rubbing some part of the child's body for a greater or less period. But this difficulty may reflect a lack of sense, or a lack of stability and perseverance, or an unhealthy emotional indulgence on the part of the parent just as truly as an untrainability or some other defect in the child.

The important thing is to maintain a healthy balance between an opportunity for the child for spontaneous development on the one hand, and intelligent guidance of the development of his potentialities with a clear vision of what good individual development is, on the other hand.

As regards the element of parental emotional indulgence as a possible cause of the untrainability of the child a good deal might be said. The study of the adult neurotic, as well as of the maladjusted child and his immediate setting, has brought to light many facts to justify the conclusion that parents tend to make the most of the natural satisfaction which the loving care of a baby brings with it. Parents do frequently express themselves as "hating to see their children grow up." An exaggerated tendency of this kind, especially where the need for this satisfaction is kept alive because of a starved love-life of the parent, may do the child a good deal of harm in making it difficult for him to acquire those habits of adjustment to the pleasure-pain elements of his life which are absolutely essential for healthy adulthood.

But in the training of the child it is important to appreciate what Professor Jennings[3] terms "the rule of the gradual and spontaneous development of the powers." He says "much of the power gained by the young human being as the years pass is not brought to him primarily by training, by learning, by the exercise of the particular faculty involved, but is a mere consequence of unhindered healthy development. After the child reaches a certain stage of development, it can do easily and quickly what it could not do even with much training at an earlier stage; and this silent unfolding may and should continue throughout life." Training is even harmful when it comes earlier than the development of the power which it tries to train. It is therefore folly to expect such functions as careful control of the bladder, or sitting nicely still, or concentrating, before the powers which make these functions possible have developed.

While we are at this point, it might not be amiss to refer briefly to some of the childhood manifestations which persist as troublesome and disconcerning habits beyond the period ordinarily considered natural to them. We have in mind such manifestations as bed-wetting and soiling of clothes, thumb-sucking, peculiar notions as regards food with a tendency to aversions which may lead to vomiting, a general babyishness of demeanor in speech and action. One should not class these manifestations with the more serious neurotic disturbances of childhood, such as emotional tantrums and explosions, night terrors, convulsive manifestations and the like. One deals here primarily with an unhealthy persistence of phenomena which are more or less natural to a certain period of childhood. Their unusualness consists in being out of tune with the setting, out of time and place.

Now in considering these conditions, one meets with a situation similar to that met with in other manifestations of conduct, namely a multiplicity of causes, and it is thoroughly safe to distrust any remedy which promises to be a cure-all for all cases of bed-wetting, for instance. Let us consider this manifestation for a moment.

In the first place we look upon the child structurally. He may have an abnormally small bladder and not be able to retain the contents for the required time to avoid wetting the bed. Or he may be suffering from excessive irritability of the neck of the bladder and thus find it difficult to control his sphincter muscles. Or the lack of sphincter control may be due to more obscure causes which are in some way related to an endocrinologic unbalance. At any rate, attention to this side of the child seems to bring about not only a better control of the involuntary musculature but also a greater capacity for the exercise of the will generally. These physical and physiologic causes must be eliminated before we come to consider strictly psychologic ones. And here the situation may be due, especially in younger children, to faulty habits in connection with these functions.

Occasionally the manifestation becomes, somehow, associated with a pleasurable organic feeling not unlike sexual excitation. Cases have been known where bed-wetting came to serve as a substitute for masturbatory activities. But it need not surprise us to discover in some instances a more positive meaning in this disorder in the nature of a mechanism for the control of the environment. I recently had a boy of 11 under my care suffering from this difficulty, who managed to control it for several weeks on a stretch during a certain period when he was paid a dollar by his father for every week that passed without an accident. Bed-wetting as a definite weapon of retaliation against a hostile environment has also been known, and it is surprising that some correctional institutions for children do not have real epidemics of this disorder more often.

In connection with the question of local irritation, I wish to bring to your attention what has been termed not very happily, the "rule of attention," which is to the effect that throughout development and all activity, only one thing can be well attended to at a time. This means that the child whose attention is claimed by some irritating bodily condition cannot give his full attention to other matters at the same time. This principle is of considerable importance, especially in later child life in connection with school failures which owe their chief cause to a distressing or disturbing content of the child's mind. Even where this disturbing content does not create a "conflict" of any intensity, as happens, for instance, when the child becomes self-accusatory, or fearful of some dire consequences to follow some mischievous indulgence, the mental content may so claim the attention of the child as to seriously interfere with his capacity to attend to changing requirements of his environment. Therein lies the great benefit in cultivating and holding the child's confidence on the part of at least some one adult in the household. This gives the youngster a chance to unburden himself of the distressing mental content, and particularly to get a wise, objective explanation of things which have been troubling him merely because they were unsolved mysteries to him.

As regards thumb-sucking, there seems to be very little doubt as to its pleasurable possibilities for the child of a kind which are closely allied to erotic sensations. The persistence of this childhood indulgence, as is well known, can be frequently traced to the pernicious pacifier. Of considerable interest and practical value is the fact that these various manifestations of holdovers from infancy seem to be linked together as parts of a more general tendency to cling to the benefits and indulgences of infancy. Thus the ridding of the child of one habit serves to check some other simultaneously existing habit. This provided, however, there is more than blind suggestion employed in curing the habit, or mere methods of harshness and intimidation. Unless the child is made to understand to the extent of his ability to do so, the meaning of an objectionable habit, the checking of one may bring out another to take its place. Thus, as we have already mentioned, unintelligent checking of masturbation may bring out bed-wetting as a substitute.

To return to a discussion of early infancy, it is wise to habituate the child to a regular and matter-of-fact method of attending to the emptying of the bowels. Many obstinate cases of constipation can be traced to an unintelligent fussing and pre-occupation on the part of the parent in connection with this function of the baby, which at times reaches the degree of a regular ceremonial.

The deep roots in a pleasure-motive which this function may have is illustrated in the many adult frank or concealed anal-erotics. It may be the root of an adult's sexual perversion. Children derive a peculiar satisfaction from messing with filth, an indulgence which is ordinarily outgrown, but which leaves in some individuals live elements for a future neurosis. A common-sense attitude should be cultivated toward the question of cleanliness, not merely because it is nice and pleasant to the parent for the baby to be clean, but because it is essential for healthy bodily growth to maturity, an appeal which carries weight with most youngsters.

The various physical diseases of infancy and childhood may, of course, leave very serious after-effects which interfere with normal development of the personality. Particularly is this true of those affections which involve the nervous system, as meningitis, for instance. Aside from its possibility of causing a serious arrest of mental development even to the degree of idiocy, it may be responsible for a general nervous instability or hypersensitiveness which hampers the child in all sorts of directions. It is important to search the developmental history of the individual we are studying for events of this nature, the after-effects of which are frequently overlooked. The child's manner of reacting to an illness may serve as an index to his nervous organization. Some children react with a high fever to every upset in their physical equilibrium, while others are stolid in the face of these encroaches. Some children readily become delirious, or very fretful and restless in connection with physical disorders. From the point of view of psychopathology it is of utmost importance to note the manner in which the adults surrounding the child were in the habit of reacting to his illnesses. If the child's illness served to throw those about him into a panic, as it not infrequently happens, especially with first babies, the child will sense this sooner or later and is apt to resort to it in case of need for throwing the environment into a panicky state. Many a child has been heard to exclaim when he found himself balked by the elders of the household, "I know what I'm going to do, I'm going to get sick, or I'm going to vomit again." On the other hand, the over-emphasis of a child's illnesses in his presence, or the dwelling upon these events by an over-solicitous parent through a constant reiteration of the seriousness or magnitude of the poor youngster's illnesses may instill in the child a feeling of being different from other children because of his various illnesses. He may make a virtue of this, especially if his mother's solicitude serves also to protect him from his sisters and brothers or playmates and gains for him a preferential position in the family. Or he may come to consider it as a disability and because of it will shrink from the ordinary pursuits of childhood and from the multitude of chances which every youngster must take in testing out the world with its possibilities.

In either case, the manner of the parental behavior in connection with the child's illnesses, especially if these events happen to be frequent in his early career, may condition more or less permanently the individual's later behavior when he has to meet life as it is in reality.

These patterns of behavior which we all carry with us from childhood in one form or another come about in the manner of the conditioned reflex and are sometimes very difficult to modify. The child comes to associate repeatedly a certain situation in his environment, a certain reaction on the part of his parents with the event of his illness. He becomes aware in connection with these events of certain special benefits with the pleasurable feelings that go with this, or when the reverse is the case, of a feeling of discouragement or defeat which is bound to lead him to under-estimate his personal worth.

In any event, these experiences of childhood illness, which are always occasions of special significance in the household, have the capacity of determining the future adult's personal morale or sense of self-esteem, a healthy possession of which is so essential for adequate adaptation.

But the child is also an exquisitely imitative creature and its development may be profoundly affected by the behavior of the parents with reference to their own illnesses. Compare the possible effect upon the child of the continuous presence of a mother or father who is always suffering from one sort of illness or another, with the inevitable, "You're making mother nervous," or "Don't play so hard, mother has a splitting headache," etc., with the influence upon the child of a cheerful, hearty parent, who appreciates that a splitting headache may be the symbol of a weakness which one should keep from the children as well as a certificate entitling the holder to special consideration and a right to domination through weakness.

One is not of course bent here upon discovering some magic process for the elimination of illness in parents, or the growing frequency of chronic minor invalidism among the child-bearing members of the human species. But this is true, that frequently the nervous, maladjusted, weakling of a youngster, or the equally maladjusted young blusterer and bully, cannot be helped very much until the parent whom he is imitating quite unconsciously in his own way, is dealt with therapeutically. It is the recognition of the imitativeness of the child and of the frequency with which the child's maladjustment or misconduct merely mirrors some unhealthy aspect of the mental hygiene of the adults who surround him that has made us more cautious in attributing so much of the evils of mankind to heredity.

In the introduction to this chapter we have already referred to the ease with which purely environmental influences in the child-parent relationships might be mistaken for hereditary effects. This is particularly true concerning the affective relationships of the child already touched upon in the chapter dealing with the sexual life of the child. But here again, the potential dangers which are contained in a too deep affection between members of a family should not lead us to the counsel of an avoidance of these love relationships. Love between parents and children is one of the most powerful influences for good in the bringing up of the child. Love and sympathetic understanding are indispensable aids in the growth from dependent childhood to a healthy maturity. This is well illustrated in the pernicious effects so frequently met with in connection with the broken-up home and the necessity for institutional rearing of children. Forty-two per cent. of repeated offenders at Sing Sing prison had spent their childhod in institutions. But we have come to recognize the occasional necessity, for obvious reasons, of deliberately breaking up some homes if the children of the household are to be given a fair chance for healthy development. Equally essential is it to recognize that this most potent factor in civilized life, the family, carries within it hidden elements of danger to the individual, potentialities for its own disruption by rendering the individual unfit for family life.

Now illness of the parent, especially a chronic state of ill health which demands for the sufferer constant special attention and concessions, may be the starting point for an unhealthy child-parent relationship of the kind we have spoken of.

Thus far we have discussed issues in child life which are more or less closely related to his existence and development as an organic being.

There soon comes a time, somewhere between three and five years of age, when more or less deliberate steps have to be taken for the direction and guidance of the growing individual, especially as regards his relationship to others. The child is coming to be increasingly confronted with the problem of social integration, of life as a socialized being and an increasing number of concrete prohibitions and denials form part of his daily regime.

Now the character of the personalities who are to impose these prohibitions, the manner in which they are brought to the child's attention and the degree of coöperative insight it is possible to maintain between the adult and the child, determine to a very large extent the manner in which the child will take in these prohibitions and the nature of his reaction to them.

This is not the place for a discussion of the natural or God-given rights and privileges that go with parenthood. In practice, that parent maintains the respect and obedience of his children, who has been fortunate enough to earn it. A child may be made to conform to certain standard formalities of child-parent relationship, but the true value of this formal behavior depends entirely upon the spirit back of it.

Neither is this the place for a detailed discussion of the rules governing habit formation. But the situation of training a child in good habits by a parent or teacher is not quite analogous to that met with in the objective laboratory study of habit formation. The difference is due to the very important element of the "exercise of will" on the part of an adult in connection with the desire to influence the child in the right kind of behavior. The element of incentive which is of such importance in this connection may be interfered with seriously when it becomes altogether too much identified with the personal wish of the trainer. A child may well wish to do a certain thing because "mother wants you to do so" or because "teacher says so," but this is a very uncertain incentive since it depends so much on the child's emotional attitude towards the parent or teacher. It may be just as strong an incentive for the child not to do as requested as to do so. It is important, therefore, whenever possible, to give the child a reason for the desirability or undesirability of a given act in language which he understands, and when illustrations are employed it is desirable to take these from the experiences of child-life and not from those of the adult. The child's preference for guidance and instruction from his peers, from his playmates and members of the gang is undoubtedly related to the fact that the adult's guidance is so rarely free from elements of coercion and compulsion which to the child may mean in his own way nothing more than the exploitation of a natural advantage on the part of the adult.

Where this element is carried to extremes, especially when the child is burdened with one of the "mighty fathers" of the human race, it is apt to influence very decidedly the future character and behavior of the individual. Here as in connection with our discussion of unhealthy affective relationships between parents and children, it is the very constitution and nature of man which exposes him to an unhealthy exhibition of parental authority. It would lead us altogether too far afield to enter here into a detailed discussion of the purely affective reasons for a parent's difficulty to get along with his children. Flügel[4] has discussed this problem very thoroughly in a recent book. To the helpless infant, the mother or nurse who cares for it and feeds it, is the symbol of peace and comfort, whereas the gruff-voiced, awkward father is from the beginning something to fear and to be suspicious of. Only gradually, and very much later than mothers, do fathers get acquainted with their children. In modern civilization particularly, the limited and sometimes infrequent opportunity which the father has for close contact with his household, may mean for the child chiefly an occasion for a scolding or correction, for which the mother has in many instances already prepared him. It is of relative import in this connection that fathers so often disappoint their wives and violate one of the most important principles of discipline, namely the necessity for a close association of reward and punishment with the event that calls for it. The repeated association of father with corrective procedure, the memories of these events as well as of those occasions when mother had to intervene as the rescuing angel, are apt to cultivate in the child a thoroughly unhealthy attitude towards sources of authority in general, and to affect deleteriously his broader relations to life in general.

Where paternal authority is carried too far, especially when little or no attempt is made to justify in the child's eye this authoritative attitude on grounds other than the natural relative position of father and child, it is apt to provoke reactions which may have a two-fold, equally disastrous effect upon the personality.

We see all about us individuals, not only those suffering from frank nervous manifestation, but others whom we would not class as neurotic but who are nevertheless seriously hampered in their behavior by an old fear and intimidation which they have carried away from their childhood relation to their fathers. They need, throughout life, the guidance and support of some father, of some person in authority or of some authoritative (so-called) opinion, and find it very difficult to form independent judgments. The success of the political boss, of the type of labor-ruler with whom we were recently made acquainted through the press, of the various charlatans who can always gain a following in a democratic country, is made possible to a very large extent by the large numbers of intimidated adults who must still have a father to guide them.

On the other hand, for reasons either of personal make-up or of special environmental circumstances paternal authority carried to excess may bring forth in the child an intense and permanent dislike of all authority no matter in what form it is met with in later life. We are all familiar with the chronic protestant and rebel to whom regulation of any sort, whether in the form of religion, custom or law is extremely distasteful and who quite blindly and almost automatically disagrees with you as soon as you have expressed an opinion. It is the growing belief of students of the personality of the chronic offender against the law that in certain instances one meets with in these individuals a life pre-occupation with the task of getting even with authority. It is true that some of our institutions for juvenile delinquents and some of the processes of the criminal law contain real possibilities for instilling in an individual what has been termed an "anti-social" grudge. But the person who has lived a normal childhood is apt to have acquired very substantial mechanisms for the avoidance of an unhealthy sense of dependence upon the opinion of others as well as for meeting those regulative requirements of social-adaptation which are part and parcel of life in a community, objectively, and without personal bias.

An important element in the difficulties which people have in assuming a healthy and workable attitude towards the question of authority, a question which every individual must face, is the element of insight. The roots for one's biased attitude or opinion may lie outside of the individual's knowledge. He may be impelled here as is true of other aspects of human behavior by unconscious motives. Once, however, an opinion is held or an attitude adopted, it is difficult for the average individual to escape the tendency to bolster it up, since it has become an attribute of the ego and as such it has to be defended. This process of defense has been termed "rationalization."

"Our rationalizations," as Wells[5] puts it, "give a motive which our personality will accept as a fitting one without giving the real cause of our actions. Rationalizations play their special parts in justifying something we are not really sure of, and in bolstering up an impulse against counter impulses that are themselves strong enough to block it." "Who then tries to be true to his rationalizations is a traitor to himself. The test of character is the abiding by form-standards of certain things we will do and certain things we will not do in defiance of the rationalizations that come every day to help us tear them down. The ability to rationalize an act is small guarantee of its real moral or logical value."

This much for the rôle which affective and authoritative relationships between parents and children play in the shaping of personality. Other influences which may have an equally determining effect come from the child's brothers and sisters and his playmates. In all these relationships we must view the child as a striving, living being, impelled by his instinctive trends to make an ever wider contact with the environment, to expand and enlarge his ego, but also to want to be like others. Regulative approaches therefore need not meet with universal refusal and opposition on the part of the child provided the manner of this approach does not elicit from him an untoward reaction. The child's reaction to the environmental stimuli, spontaneous or those deliberately shaped, is the "criterion" in the last analysis of the value of these stimuli for that particular child. The child who misses in his home relationships contact with adults whom he can love and admire is deprived of one of the most helpful vehicles which make possible growth from an ego-centric, asocial being to mature, socialized adulthood. Moreover, the reaction may be of a more positively injurious kind since by making difficult normal growth, it renders the various self-loving tendencies which make up the effortless pleasure-pain existence of infancy doubly attractive to the child, making it harder for him to cultivate those reactions which are necessary for meeting the realities of life.

In other words, an unhealthy mental hygiene in the development of a given individual may be due not only to bad training, but to the deprivation of the child of certain natural needs, to the thwarting of a certain natural craving which makes good training impossible.

It is probably true that the only child is exposed to greater obstacles to normal development than the child who grows up in the company of brothers and sisters, provided there is not too great a gap between the ages. At any rate a large percentage of adult neurotics are found to have been only children. The reason for this does not lie solely in the fact that an only child is apt to be spoiled by his parents in the conventional sense of the term, but because he is deprived of the opportunity for free and intimate association with other children, an experience which contributes normally very decidedly to the process of growing up. It is in connection with the brothers and sisters that the child first comes upon the necessity for taking cognizance of the wishes and interests of other persons. It is in the group activities of the nursery and playground that the first contacts are made with the game of life, and the things which the child has failed to learn in the nursery he has to learn at a later day and at the expense of much greater effort and, perhaps, suffering.

But here, too, contact with the personalities of other children, especially with brothers and sisters, because they are praised and blamed by the same parents and are therefore contenders on the same field of battle, carries with it certain dangers along with the benefits. The relationship is not always or even commonly one of love and mutual coöperation. On the contrary, frequently enough the relationship between little brothers and sisters is tinged with considerable jealousy and hate. This is especially apt to be the case when an unhealthy competitive spirit is encouraged, or when the occasions for deep feelings of defeat or victory are supplied by the parents through an undue stressing of traits of character of which the child is made to feel unduly proud or ashamed. The situation here is similar, though perhaps more significant because it concerns brothers and sisters, to that met with in the classroom where one or two pupils are singled out from all the others as deserving special recognition in the form of a prize. Unless all children of a household are made to feel in one way or another that they have the esteem and love of their parents, attitudes are apt to be cultivated on the part of the neglected ones which are very harmful to the personality.

The wise parent will see to it that in so far as his affective relations with his children are concerned, each one has a fair and equal chance, and although he will usually succumb to the inevitable tendency to have a favorite, he will see to it that neither the one so favored (or cursed) nor the others will become aware of this.

In this connection reference might be made to the almost universal tendency of parents to shape the ambitions of their children. Now there is nothing wrong about this just so these ambitions are based upon a clear recognition of the physical capacities, natural inclinations, and intellectual and emotional equipment of the child, and not solely upon the hope of gratifying a wish of the parent. Altogether too often the parent is moved in this connection by a desire to make good some failure or disappointment of his own, to seek compensation in the life of his offspring for a personal deprivation.

The world is full of people who are struggling with a conflict which is related to an unbridgeable gap between aspiration and native equipment. The wisest course to pursue in this regard is to give the child first of all a chance to show his native capacities so that he may know himself, give him an opportunity to develop his capacities, then let him choose. Even expert advice in vocational guidance, such as is possible with our present state of knowledge, might then also become a matter of his own seeking, so that whatever rôle he is ultimately destined to play in life will be something in the shaping of which he participated and for which he can assume personal responsibility. This does not mean a disinterested attitude on the part of the parent toward his child's choice of a life career. Neither does it mean a negative policy in the matter of cultivating proper ideals in one's private and business life. What is meant is the avoidance of the pernicious tendency of squeezing a child into a mold cast for him by his parents whether or not he is meant to fit it.

Some consideration of the very important aspects of the child's life up to the age of five, which are related to the development of his sex instincts, cannot be omitted here. It is in connection with the child's recognition of the earliest stirrings of this instinct that the attitude of the adults about him assumes tremendous significance for good or evil. One's later tendency to view the phenomena of life and one's relation to others in a frank, natural, matter of fact fashion or to meet life's issues in a sensitive, shut-in, awkward manner may be conditioned by the manner in which the child's first expressions of curiosity concerning his sex stirrings were met by those to whom he addressed himself.

It is remarkable how readily the general secretiveness and inaccessibility of a maladjusted and troubled youngster may be dissipated as soon as he has talked himself out freely concerning his sex curiosities or troubles. On the other hand we see extreme results of an unsatisfied curiosity along these lines or of an induced secretiveness because of the manner in which some adult has met the child's natural and innocent curiosity along these lines, in various forms of neurotic manifestations and family maladjustment. Occasionally it seems to be the central factor in the development of a grave mental disorder commonly known as dementia precox. Ordinarily the adult whose own sexual life is managed in a healthy manner will find no difficulty in answering the child's innocent queries in a natural manner. Certainly there can be no doubt concerning the innocence of the child's inquiries. Whatever other characteristics they may impress us with, we usually read into them, in accordance with our own attitude toward sex.

And yet of all the positive mental hygiene principles we know of, this, the cultivation of a frank, natural and clear-sighted attitude of mind is one of the most important for healthy adaptation.

With the advent of school life, new exposures to influences, good and bad, come into the life of the child which have a decided influence upon the shaping of his character. A good many of the issues in this period of life which deserve our attention fall strictly within the field of pedagogy and cannot be considered here. But one of the outstanding elements of the present-day agitation for reform in the educational field is intimately related to our own field of mental hygiene. I refer to the growing insistence upon the recognition of the child as a living and striving human being with a variety of interests, a variety of inclinations, a variety of potentialities and not merely as a receptacle to be filled up with a prescribed assortment of information. A still further extension of this principle would demand for the child the opportunity in connection with his school career for a type of personality-expression which means more than mere scholastic performance. Along with this should go the right to be judged and estimated by his elders as a human being with all sorts of desirable and undesirable qualities and not merely as a scholastic performer. Education and such knowledge as the formal educational process gives the individual is a powerful weapon in the task of adaptation, but it may actually prove worse than useless if it is not accompanied by some knowledge in the business of living.

It is evident that the first step in the modification of the traditional attitude of formal educators toward the child is a more comprehensive view of his origin, his developmental history, of the type of experiences which have come his way during his first years of life and of his general reactive tendencies, at least. This involves a much better coöperation between the home and the school than was required when the child was consciously or unconsciously presumed to bring with him to school only that part of him which characterized him as a school child.

This is being achieved to a very large extent already with the maladjusted child through the medium of the visiting teacher and other similar social-educational enterprises. But the time is bound to come when every child upon entering school will receive a well-rounded study of his personality such as we have referred to.

A great deal of progress has already been made in the direction of obtaining a dependable estimate of the child's intellectual equipment and in the matter of fitting his educational program to this equipment.

Not enough has been said in emphasis of the emotional impediments to the proper use of whatever equipment a child may possess.

We have had cases of school failure in children with high intelligence quotients, sometimes with exceptionally superior intellectual endowments, whose failure to meet the requirements of the curriculum could in no way be explained on the basis of lack of equipment. Moreover, we have seen children in whom a frank stunting of such abilities as they did possess was brought about through wrong educational procedure. But we shall not concern ourselves here with matters of strict pedagogy. From the point of view of mental hygiene, education means the "fitting of the individual for the business of living," a very important element of which is the cultivation of proper methods of using one's mind and personality in the task of adaptation and the correction of unhealthy adaptive dispositions.

The educator's very extensive contact with the developing individual during the period of school life puts an obligation upon him and offers him the opportunity to concern himself with these aspects of education, and the manner in which he expresses this concern will be indicated by the answers to the following questions, which, according to Dr. Adolph Meyer,[6] we might ask concerning a school system:

1. Can the result be called well-rounded, making for preparedness, for an efficient and wholesome 30 or 40 or more years of adult life?
2. Does the product of such training know what he or she is fit for; what he or she wants or will try to do as his or her share of the work of sustenance and productiveness?
3. Is the school training in harmony with our best knowledge of the integrated human organism and personality?
4. Does it satisfy the principle that life ultimately be judged in terms of a biography of objective achievement, and that knowledge is merely an incidental asset and telling only when it shows in effective or expressive activity?
5. Does it succeed in forging the natural assets into power?
6. And finally, what share can any type of school have in favoring or not favoring the individual chances for mental and moral health and efficiency?

Concerning the first question, it may be said that whatever will ultimately come to be considered a well-rounded education will have to embrace as one important element, a thorough knowledge of the human machine and of at least some of the common fundamental principles of the hygiene of the mind.

Among these, what Professor Dearborn[7] considers "mental economy" might be placed first. "True mental economy is a distributive virtue and consists not in saving but in selection." One should choose the things which one believes most worth while, and if he is to enjoy the best of mental health and happiness these must also be the things which he most desires to do. Mental stress and strain begin when one's work is not the expression of his desires but conflicts with them.

Another fundamental principle which ought to be included in a well-rounded education is the necessity of acquiring the habit of concentrating attention on the one thing in hand, of learning to live one day at a time. So important is this for the mental health that one's ability to concentrate upon the present, ignoring the past and the future except as vitally related to the present, is in a certain sense, a gauge of one's sanity. To be single-minded and undistracted through the course of actions of a life-time, the student must have discovered some permanent interests and aptitudes. The reverse of this is observed in individual's who have fallen into the unfortunate habit of doing everything at least three times or at any rate of wearing themselves out about having to do a certain thing, in worrying while they are actually doing the thing, and worrying all over again about the way in which they did it. In connection with this, it is of great value to cultivate the habit of "doing the best thing" in a difficult situation and let it go at that.

The craving for an omniscience and omnipotence in the manner of meeting life's problem is a vestige of the compensatory day-dreams of childhood dependency and can never quite be attained in real life. This recalls another principle, namely, that "the laws of mental health and character require the completion of thought or feeling by expression in action." Mere feeling and thought and fancy which are not brought to the test of action, to their fulfillment in action, tend to become one of the danger points of human nature. Especially is this true of that form of adaptation which works through the "imagining of a realization" of a thing wished for, which through lack of strength or of a unified will cannot be attained directly. The day-dream is a very common example of this. Here a genuine pleasure is experienced from picturing the dream as realized though the sense is preserved that it is not so, and one does not act as if it were. In mental disease, however, these fancies seem real and the individual acts upon them as though they were real.

In so far as the educational process undertakes the training of the student in the use of his mind, it ought to aim to habituate him to "orderly association of thought." "Disorderly association means the beginning of conflict and worry." In the course of educating the child, tasks should be given which are simple and definite, instructions clear and concrete; decisions and actions should be straightforward and whole-hearted.

On the other hand, school life may be anything but a beneficial experience for the individual. The tasks imposed by the educational process may prove much of a strain upon the nervous organization of some children, especially when the child is being driven beyond the means of endurance by over-amibitious parents. Cases of this kind have come to our attention.

The reverse situation maintains when the unusually capable and well-endowed youngster is kept at tasks which do not call forth all his capabilities, bore him to death, allow him to fall into bad habits of concentration and application or finally make him detest the boredom of school life. From the point of view of psychopathology the psychology of success and failure in connection with school life is of utmost importance. A chronic state of defeat, or the persistent lack of the stimulus of achievement tend to hamper the child in the use of those capacities which he does possess, and some day more will be said by the psychologists concerning "functional feeblemindedness."

One other point might be mentioned in connection with the mental hygiene of the school period and that is the necessity for coöperation between school authorities and parents in the matter of discipline. A teacher's work of weeks in connection with the training of the youngster in some desirable personality attribute may be undone entirely through lack of coöperation of the parents, especially if in some way the child is given to understand by his parents that they do not hold the teacher in high esteem. Parents do not realize how their own management of some of the child's school difficulties puts obstacles in the way of achieving the very thing they are after.

Our discussion of the school-life of the child is perhaps more detailed than is ordinarily accorded this subject in medical books, but it finds its justification in the growing demands that are being made upon the psychopathologist for advice in connection with child rearing. The physician who would limit his interest in connection with a maladjusted child to the strictly medical issues will frequently fail to help the child, and when his own equipment does not embrace some knowledge concerning educational-psychology, he will do well to procure this advice for the child from the proper sources.

The period of childhood, psychologically considered, might be said to terminate at the age of 12, though certain of the elements in child-life which are of considerable importance psychopathologically, merge imperceptibly with the period of puberty, a subject which deserves at least some consideration here.

Puberty ordinarily sets in in boys at 15 and in girls at 14. There are significant variations in this, depending upon climate and race particularly. But the period shortly preceding puberty and all the way on through puberty and the years of early adolescence is one of the significant periods from the point of view of personality organization and development. It is due primarily to the maturing of the sex glands with the physiologic capacity for reproduction though the individual may not be capable physically of child bearing. In connection with this a profound disarrangement of the child's mental metabolism sets in. The development of secondary sex characteristics shows itself by rapid and frequently uneven growth of body, a general instability, and vague longing and restlessness on the part of both boys and girls. The expression of such mental traits reflect a vague consciousness on the part of the individual that he has arrived at the stage of his development when he is really a member of the human family. These traits, particularly the commonly observed over-conscientiousness and auto-criticism, the romanticism and hero-worship, the tentative toying with the questions of the meaning of life, etc. may, under certain conditions, as for instance in the presence of a conflict about sex matters, or religious doubtings, or a re-estimation of the worth of one's parents, lead to the development of permanently injurious traits of character and under particularly stressful situations, to suicide.

The melancholy mood is not an uncommon manifestation in early adolescents and while ordinarily one pays but slight attention to it, we must not forget that before the war, at any rate, suicide among children had become alarmingly frequent.

The boy is just itching to test himself in the game of life at as many points as he possibly can, and longs for new experience, adventure, travel. The ordinary duties of daily routine are apt to become monotonous, irksome and very wearing to the nervous system, the general resistance of the organism is lowered and disease is very common at this period, and it is essential to keep in mind the natural reasons for these various changes and the transitory character of them in dealing with the behavior problems of the adolescent.

Sex stirring and pre-occupations which are commonly in abeyance following the early vague interests of the child up to the age of five or six, are apt to assert themselves now with considerable force, there is commonly a return to onanistic interests, but these bring with them an intense moral conflict because of the accompanying intellectual and affective characteristics of this period.

The girl is apt to form deep affective attachments to other girls, particularly older women, older schoolmates and teachers. At the same time they long for recognition as women from the opposite sex and readily identify themselves with the heroines of the novels devoured at this period. The various forms of misconduct due chiefly to adolescent instability are unfortunately still being handled to a very large degree as though they were manifestations of a permanent and fixed trait, so that the very corrective procedure employed may do more harm than good. We have in mind particularly the repressive procedure of institutional life.

Similarly, school systems do not take sufficient cognizance of the increased burden which the educational task comes to be with the onset of puberty and adolescence. No allowance is made for the diminished capacity for organized effort in the adolescent. On the contrary school is ordinarily made more difficult at this period. Yet defeats and failures experienced at this time may leave a lasting deleterious effect upon the boy or girl.

Those children who are severely tainted by heredity are apt to show manifestations of this burden at this time of life. A number of the actual mental disorders of later life may make their first appearance in mild signs of deviation from the normal at this time. Thus dementia precox and manic-depressive tendencies may appear, epilepsy and hysterical reactions, mild states of anxiety, hypochondria and melancholy may color the adolescent picture.

Particularly deserving of attention is a tendency to shyness and undue awkwardness in one's relation with others. A preference for solitude, a lack of frankness and naturalness in intercourse with others, an undue self-satisfaction in inner broodings, ruminations and day-dreams and the absence of an active, healthy interest in objectivity. These shut-in creatures, who always seem "tied in a knot," neither cooled nor warmed by the events of ordinary life, without lively enthusiasm or interest in anything in particular, are very apt to develop gradually a serious mental disease, if indeed they are not already in the midst of it.

Active attention at this time in the nature of a spirited effort to draw them out, to get them attached to reality, to ween them from their preferences for a pre-occupation with their day-dreams may check the pernicious process. Peculiar food fads, or over-scrupulousness of a religious kind, the expression of notions about queer bodily sensations also deserve serious attention.

As adolescence advances the question of companionship, of creative group interests, of opportunities for self-expression in more socialized directions become of paramount importance. It is during this period, especially in boys, that certain concrete environmental dangers must be kept in mind. Excessive smoking or experimentation with intoxicants or habit forming drugs, sex exposures which may bring with them lasting effects in the shape of a syphilitic or gonorrheal infection, intense love-fascinations which may wreck a career, and exposure to various perversities. The boy between 15 and 23 is in a mood to "try anything once" and while experience is a very good schooling for life, the dangers that lurk about this adventurous age must be brought to the adolescent's attention.

I want to mention particularly the danger of converting an unfortunate sex experience of an adolescent girl into a permanent disability or a life of prostitution through unintelligent or sometimes vicious management of the situation on the part of relatives or legal authorities.

There comes a time in the life of every individual man and woman when he must achieve a proper and thorough emancipation from parental dependence and attachment. This does not mean, of course, that family ties are to be broken up or that there must necessarily be a physical separation from the parental home. But in order that the individual be able to stand squarely on his own feet, to make his own decisions and carry the responsibilities that go with this, he must free himself from the tendency of childhood and adolescence when decisions are largely made for him and responsibility shared with him by someone else.

The natural tendency under normal conditions is for this to take place as part of growth to maturity. But some parents, especially again parents who find themselves balked in the expression of their own life strivings, find it difficult to abandon their hold upon their children and hamper them in the process of full development. It is a fact, not uncommonly demonstrated in the lives of maladjusted humans that the individual who has not achieved this type of thorough-going emotional emancipation is not apt to make a successful life-mate, since his allegiance will always be a divided one, and life a continuous series of conflicts as to what stand to take. Moreover the parent who has to rush to his own parents for guidance and for assistance in making decisions in not apt to prove a very helpful guide to his own children.

In discussing the child-parent relationship of an earlier period in life we indicated the general principles which are apt to render the process of emancipation easier both for parent and child, and in cases where this is more or less of a tragedy to one or both partners to the enterprise, the causes lie very deeply in the infancy and childhood of the individual's career.

Another point in connection with this epoch which deserves mention is the question of choice of occupation. I do not mean by this the sporadic experimental work history of the boy or girl of early adolescence, which, though flighty and changeable, carries with it a good opportunity for the kind of adventurous experimentation which is very healthy, indeed. I am not entirely in sympathy with the kind of "vocational guidance" movement which would undertake the casting of individuals into industrial molds almost from the cradle.

Guidance can sometimes be carried to the point of over-guidance which results in an inevitable pursuit which the individual cannot escape because in his training and development his lot has been decided for him. Thus compliance can be secured, but a compliance of this sort may rob the world of some very vital contributions which certain kinds of rebels have always made.

I am referring here to the choice of one's life work. A man's work must carry with it certain vital satisfactions which have their roots in various innate dispositions.

From what has been said, some general conception at least might be had of the many difficulties with which the path through infancy and childhood is strewn and the rôle which these difficulties might play in the maladjustment of the child. In concluding this discussion the point might be stressed again of the extreme importance in the treatment of childhood difficulties to have some estimate of the adults surrounding the child and of the circumstances of life which constitute his "setting." Without due attention to these elements, therapeutic endeavors are apt to be futile in many cases.

Our next concern will be with an attempt at an orderly formulation of the various types of reactive manifestations of the childhood personality, and with a discussion of the extent to which the psychopathologic manifestations of childhood lend themselves to formal classifications.

CLINICAL CONSIDERATIONS

We have already indicated how unjustifiable it is on the basis of our present knowledge to try to fit the manifestations of childhood psychopathology into any rigid scheme of classification. Nevertheless there is considerable value in an attempt to state these issues, tentatively at least, with some general and comprehensive outline in view. In doing so the point must not be lost sight of that in so far as any attempt at classification from the point of view of causation is concerned, the situation is seriously complicated by the fact that in the majority of instances one deals here with a multiplicity of causes which require adequate evaluation.

It will render our task easier if we begin with the more or less well-defined conditions and proceed gradually to the more obscure problems. In accordance with this, the behavior and personality difficulties of childhood may have as central causative factors in various degrees of combination with secondary etiologic issues, the following:

1. *Arrests of Intellectual Development of Various Degrees of Intensity.*—The commonly accepted manner of stating these differences in degree of defect is as follows, though this is probably not altogether expressive of the actual situation.
   - (*a*) Idiocy, in which the degree of intellectual development is equivalent to that of a normal child of two years or under, irrespective of what the chronological age of the individual may be.
   - (*b*) Imbecility, which because of the wider range of intellectual ability involved, is again subdivided into high, medium and low grade, and in which the degree of intelectual development is equivalent to that of the normal child between three and seven inclusive.
   - (*c*) Feeblemindedness proper or moronism, in which condition the arrest of development places the individual at a level of normal childhood anywhere between the ages of 8 and 12, irrespective of his chronological age.

     These issues are stated more accurately, perhaps, as will be seen in connection with our discussion of the subject of mental deficiency, in terms of the intelligence quotient, that is, the ratio of mental age to the chronological age, using the age of 16 or 15 as the upper limit at which normal intellectual development is supposed to be complete.
   - (*d*) Another group which is receiving considerable attention is the "dull normal" group with an intellectual capacity higher than that of the moron, but falling short of normality. A more critical discussion of the value of classifying mental deficiency in this manner will be undertaken in the chapter devoted to this subject.
2. *The Constitutionally Psychopathic Children.*—This refers to an increasingly diminishing group of children, owing to better understanding and more accurate diagnostic techinque, in whom there appears to be present a constitutional defect which involves primarily the instinctive-emotional organization of

the personality. Intellectual development may be normal, but owing to an imperfect organization of the personality these children give evidence of a more or less persistent difficulty of adaptation to life.

3. *The Psychoses of Childhood.*—Clear-cut cases of mental disease are rare in childhood, but when they do occur they simulate more or less closely the clinical pictures observed in adults. Those more commonly met with are:
   - (*a*) Toxic-exhaustion deliria and post febrile psychotic states.
   - (*b*) Manic-depressive states.
   - (*c*) Schizophrenic states (dementia precox).
   - (*d*) Syphilitic disorders, commonly juvenile paresis.

   In addition to the foregoing, acute psychotic reactions of a confusional-hallucinatory nature with or without profound affective disturbances are occasionally brought on by adverse environmental situations in the mentally defective and constitutionally psychopathic children.

4. *Organic Nervous Disorders of Childhood.*—Only referred to here because these are frequently accompanied by abnormal psychic states. Here we will include, provisionally at least, epilepsy.

5. *Functional Nervous Disorders of Childhood.*
   - (*a*) Psycho-neurotic reactions of a more or less well-defined nature, as hysteria, anxiety states, compulsions and phobias. The precise relation of these conditions to the psycho-neuroses of adulthood is not absolutely clear, especially as regards the etiologic moments. But Freud's contention that the adult's psychoneurosis has its roots in a more or less similar event of childhood is of significance here.
   - (*b*) Reactive manifestations of a functional nervous type and referring to any one of the various bodily functions, ordinarily described as the various symptoms of the so-called "nervous child." Here belong above all:
     1. Convulsive phenomena (non-epileptic), spasms and tics.
     2. Various forms of speech disturbance.
     3. Various forms of sleep disturbance.
     4. Infantile characteristics of function or demeanor retained beyond period normal to these, affectations, thumb-sucking, enuresis, tantrums.
     5. Kinetic disturbances, usually in the form of a chronic, hyperactive, restless state, with a diminished degree of fatiguability.

     The most that can be said concerning these manifestations at this time in any endeavor to clarify the rather vague notion of the "nervous child," is to consider them as reactive manifestations of the personality as a whole, which are symbolized by any one or a combination of several of the symptoms described above.

6. Conduct disorders of childhood accompanied by various unhealthy or anti-social traits as reactive manifestations of a functional maladjustment between the individual and his environment.

   It is particularly with reference to this group that the issues are not altogether clear. In the first place, it is quite impossible to estimate accurately to what extent a possible constitutional factor may be at work here, even though evidence is lacking of the existence of any of the clearly constitutional handicaps already referred to. In the second place, some of these disorders of conduct are so profound and persistent and lead to so much conflict with the environment that one is not capable of venturing any predictions as to the possible element of progression. A more complete discussion of this subject will be given further on.

7. *Endocrinopathies of Childhood.*—This is perhaps the least clear of the issues to be dealt with in childhood, irrespective of the fact that in cretinism and mongolism we are dealing with more or less clear-cut entities. But the importance of the endocrinopathies goes considerably beyond these conditions and embraces a very wide field of needed research into the relation of the glands of internal secretion to health and conduct.

To what extent this classification will stand the test of a more critical experience in dealing with the problems of childhood, we do not venture to predict. But it should serve as an aid to a general orientation in this field. Some of the conditions referred to have already been discussed in these volumes and will be omitted here.

THE ARRESTS OF MENTAL DEVELOPMENT

Among the constitutional handicaps of childhood, those relating to the various degrees of intellectual defect have received extensive attention in recent years.

The impetus for this pre-occupation with the subject came to a very considerable extent from sources other than medical, as a by-product of the spirited social efforts of the past two decades or so to deal more intelligently with the various so-called social evils. For some time prior to this, in fact ever since the subject of feeble-mindedness has been the object of scientific study, it was possible to demonstrate with increasing frequency that there was a real relationship between mental defect and anti-social behavior, dependency and other forms of social inadequacy.

But it was not until 1905–1906, with the introduction of the Binet-Simon Scale for measuring intelligence that the great popular interest in this subject began to develop. Unfortunately, the thoroughly undisciplined fashion in which the subject has been exploited by non-medical agencies has led to an altogether unwarranted simplification of etiologic issues and diagnostic methods on the one hand, and to an almost panicky endeavor to explain all social ills on the basis of feeblemindedness on the other.

More careful clinical investigation, of which the recent report by Potter* is an example, as well as research in the fields of pathologic anatomy and physiology justify the belief that the problem is not as simple a one as one would be led to believe from the restriction of the field to the limits set for it by certain psychologists and that significant modifications are in order of even very recently held views of the relation of feeble-mindedness, for instance, to criminality. In the main, however, the misconceptions have been in the nature of exaggerations, rather than of fundamental fault and the subject is being appraised more and more sanely.

Definition.—The best definition is probably that furnished by Tredgold, namely, "that it is a state of mental defect from birth or from an early age, due to incomplete cerebral development in consequence of which the person affected is unable to perform his duties as a member of society in the position of life to which he was born."

More specific definition is possible when one deals separately with the accepted categories of mental defect as tentatively determined by the current scales of mental measurement. Thus the idiot is an individual who has attained a mentality equivalent to that of a normal child of two or under. His performance abilities range from absolute helplessness to an ability to feed himself. But the highest grade idiot eats indiscriminately. The imbecile stops short in his development at a mental level equivalent to that of the normal child anywhere between the ages of three and seven. His capacities are considerably beyond those of the idiot, but as will be seen later, are quite insufficient to enable him to take care of himself with any degree of success.

The moron, sometimes called the feebleminded-proper, does not reach above the mental level of the average normal child of 12. It is generally conceded that any individual of 15 or over who shows according to standard scales of intelligence a retardation of three years or more might be considered feeble-minded.

Now while the methods of differentiating the various grades of mental deficiency on the basis of the intelligence level attained are very useful and practical in their employment, it would be thoroughly fallacious to assume that the scale of intelligence as determined by standard tests differentiates these individuals in all necessary respects, or furnishes even a dependable basis for the practical management of these individuals. Within a certain grade of intelligence level there are very significant variations in the personalities of the feebleminded, variations which are of utmost importance in determining the medical, educational and social needs of a given individual, even aside from the important question of special defect and special abilities which may be discovered within a given range of intelligence as determined by the measuring scale.

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* Howard W. Potter, Personality in the mental defective, with a method for its evaluation, Mental Hygiene (July, 1922).

Walter Fernald[8] has the following to say concerning this phase of the situation: "The writer once assembled in a room 252 individuals with a mental age of eight years. The individuals in this group varied in physical age from 12 to 50 years. Some of them had learned to read, while others had not been able to do so. Some were capable of elementary computations, while others found the simplest concept of number almost beyond their capacity. We had been able to develop some of them to become fairly expert mechanics, but others were able to do only the simplest sort of manual labor. Some were conscientious and relatively trustworthy; others were most untruthful, dishonest and unmoral. In some sex proclivities seemed to be the dominant interest, while in others the sex interest seemed to be entirely normal. This variability shows that a measurable intellectual level is not the only factor in the study of the feebleminded and in the working out of the type of care and training that they need.

"These variations would be understood if we should examine the brains of a group of cases with eight-year mentality. We should find that the structural lesions of the central nervous system which cause the mental defect vary as greatly as the behavior characteristics of the group. In certain cases we should find 'naked-eye' lesions caused by actual traumatism or as a result of faulty development, and in others the only lesions discoverable would be minute alterations in the ultimate structural elements, visible only with the microscope. The location of these lesions, both gross and microscopical, has also infinite variability, and consequently varying effect upon the mentality of the patient."

Moreover, it would be quite incorrect to assume that the differences between the feebleminded and the normal are purely quantitative in nature. No normal boy of eight years of age would wish to be compared with a feebleminded individual of 18 with the same mental level of development. It is not until we come to estimate the entire personality of the mentally defective, his affective-instinctive life as well as his intellectual equipment, that we appreciate the differences between the mentally defective and the normal.

A more strictly clinical approach to the problem involves a method such as was recently emphasized by Potter in an important piece of clinical research on the feebleminded. As a result of this, a guide for the study of the defective has been evolved which is bound to prove of much greater value clinically than the existing scales of intelligence measurement and which, because of the practical aid it is bound to give the physician in the management of the individual case, we reproduce here in detail. It must be remembered that the various groupings in accordance with certain outstanding traits have been arrived at as a result of actual clinical study.

He approaches the problems in the following manner;

THE GUIDE

SECTION A

Traits That in a General Way Tend to Indicate the Subject's Intellectual Characteristics

Type I. Dull, slow to learn, no spontaneous interests, judgment and planning capacity poor.

Type II. Moderately alert, learns fairly easily, no spontaneous interests, judgment and planning capacity immature.

Type III. Alert, bright, quick to learn, spontaneously interested, good judgment and planning capacity.

SECTION B

Traits That Tend to Indicate the Sense of Responsibility

Type I. Actively irresponsible, heedless, contemptuous attitude. Unconscientious, not orderly, unwilling to be helped, heedless of the rights of others, and contemptuous of the usual standards of social and economic fitness.

Type II. An undeveloped sense of responsibility. Lacking in conscientiousness and unappreciative of the rights of others. An undeveloped sense of social and economic standards. Not unwilling to be helped, but unable to help themselves in any spontaneous, consistent, constructive fashion.

Type III. Fair sense of responsibility, conscientious, orderly, appreciative of the rights of others, and realizing the necessity of conforming to social and economic standards. Willing to be helped, and ever ready to take advantage of anything in order to help and improve themselves in a constructive way.

SECTION C

Traits That Tend to Indicate the Industrial Efficiency of the Individual

Type I. Thoroughgoing, steady, and persistent in application. Requiring more or less constant direction.

Type II. Thoroughgoing, industrious, steady, and persistent in application. Requiring practically no direction.

Type III. Superficial, careless, poor in application, and unsatisfactory even with constant direction.

SECTION D

Traits That Tend to Indicate the Amount of Nervous and Muscular Energy

Type I. Slow, deliberate, unhurried, with little or no initiative.

Type II. Inactive, sluggish, desultory, lazy, no initiative, and easily fatigued.

Type III. Active, energetic, quick, consistent, considerable spontaneity and initiative.

Type IV. Variable activity—at times sluggish and indolent, at other times overactive and tense. Active by fits and starts.

SECTION E

Traits That Tend to Indicate the Types of Habitual Reaction to Subject's Inferiority

Type I. Compensation-reaction type (boastful, expansive, over-confidential, play for attention, trouble makers, sex and stealing delinquencies).

Type II. Defensive-reaction type (stubborn, unwilling to follow advice, sensitive to criticism).

Type III. Defensive-reaction type with secondary elaboration (paranoidal types, suspicious, discerning hidden motives, feelings of self-reference, mild persecutory notions).

Type IV. Evasive and neurotic-reaction type (hypochondriasis, sympathy seekers, capitalization of physical feelings and discomforts, blaming circumstances and others for own faults and mistakes).

Type V. Devoid of reactive tendencies, with the exception of lack of self-confidence.

SECTION F

Traits of Especial Significance to the Individual in His Social Adaptation, with Particular Reference to Amiability and Sociability

Type I. Amiable, friendly, sociable, facility in making friends, gregarious.
Type II. Amiable, but reserved, slow to make friends, not actively gregarious, but yet not actively solitary.
Type III. Shut in, distant, unfriendly, self-centered, and actively solitary.

SECTION G

Traits of Especial Significance to the Individual in His Social Adaptation and Particular Reference to Tendencies Socially Unfavorable and Those Clearly Anti-social

Type I. Quiet, well-conducted, well-mannered, respectful, coöperative, easy to get along with, no active delinquent tendencies.
Type II. Same as Type I, but in addition naive and easily influenced or led into sexual discrepancies, stealing, or other anti-social behavior.
Type III. Boisterous, loud, ill-mannered, difficult to manage, actively seeking delinquencies of a sexual or other nature.
Type IV. Ingratiating, cunning, deceitful, underhand, undependable, untruthful, with delinquent potentialities.

SECTION H

Traits Especially Related to the Prevailing Mood

Type I. Cheerful, and happy in a simple, quiet way.
Type II. Cheerful, and happy in an aggressive and spontaneously active way, heedless of the feelings of others and loudly and boisterously exuberant.
Type III. Phlegmatic and indifferent.
Type IV. Grave and serious, but not depressed.
Type V. Depressed, unhappy, and pessimistic.
Type VI. Variable mood. At times cheerful and happy in a spontaneous, aggressive fashion, at other times unhappy and depressed.

SECTION I

Traits of a Reactive Nature, but Closely Associated with the Prevailing Mood

Type I. Good sense of humor, even-tempered, stable, patient and complacent.
Type II. Poor sense of humor, emotionally unstable, impatient, fault-finding, irritable, quick-tempered, sullen and sulky.

SECTION J

Evaluation of Traits, Interest, and Aptitudes That, Owing to Their Intrinsic Nature, May Especially Favor Specialized Educational Efforts in Behalf of the Subject

1. From what does the subject get his deepest satisfaction?
2. What are his leading interests in work?
3. What forms of diversion, play or entertainment most appeal to him?
4. Has he any special aptitudes?
5. Is he religious?

SECTION K

Unique and Pathologic Traits

Odd or peculiar habits, mannerisms, episodic behavior, fears, doubts, compulsions, syncopes, stuttering, tics, abnormal dreams, somnambulism, delusions, and hallucinations.

Now to obtain this kind of estimate of the feebleminded individual it is necessary not only to observe him carefully in his daily conduct, but to obtain a careful and detailed account of his developmental history, of his reactions to the ordinary events of life, etc., aside from a thorough physical and mental examination.

Prevalence.—This aspect of the situation has also suffered from the unbridled enthusiasm which has surrounded the subject in the past few years. These difficulties which might render a correct estimate of the situation possible are almost insurmountable. In the first place, with the exception of the State of Massachusetts[9] there is not in operation anywhere else a system of inquiry which makes possible the detection of the feebleminded before they come in conflict in some way with the social order. It is true, sporadic surveys have been carried out here and there which give us a cross-section view of the situation at a given place and time, but what is needed is a continuous alertness in the form of a permanent machinery, probably in connection with the public school system, that would detect the mentally defective at least at the time of entrance to the school. It will then be found that the prevalence of the feebleminded in those institutions which serve for the detention of social pathologic elements of the community, that is, jails, almshouses, penitentiaries, reformatories, prisons, etc., will be very much decreased. In other words, the sooner the feebleminded are recognized for what they are, namely, problems in mental pathology, and treated accordingly, the less likely are they to become problems in social pathology. There are certain feebleminded individuals, particularly those of the imbecile grade, who are burdened with a combination of a great hypertrophy of the pugnacious instinct and lack of inhibitions which renders them a menace under all conditions.

But for the majority of the mentally defective, it can be said that the problems which they create are of a negative nature, are problems predominantly in social inadequacy and result in many instances from the unfortunate combination of feeblemindedness, unfavorable environment and unintelligent management.

The situation might be accurately described by saying that the feebleminded furnish the raw material out of which unintelligent management and social laxity create the problems of feeblemindedness. It has been repeatedly shown that the properly managed defective is not only no menace to the community but that in many instances he may be made to lead a serene, peaceful and fairly useful life. Leaving out of consideration the numerous extravagant statements that have been made concerning the prevalence of mental deficiency, it might be asserted with some degree of certainty that probably from three to four in every thousand of the population are feebleminded.

Causation.—Notwithstanding the increasing doubt which is being entertained concerning the extent of the rôle of heredity in mental deficiency, this factor must still be considered the most important single causative factor. Goddard[10] estimates that anywhere between 65 and 80 per cent. of feebleminded persons have had feebleminded ancestry. One deals here with a defective protoplasm which transmits its own limitations. Researches in calcium-metabolism and in the general field of endocrinology promise a more specific definition of the nature of this ancestral defect, but at present the situation is still very obscure. The data which have been supplied from the study of defective families such as the famous Kallikaks and Ishmaels are not altogether convincing and cases come increasingly to light of feebleminded individuals begetting normal children. Other causes are traumatism, especially at time of birth, toxic agents such as syphilis and alcohol in the parents and some of the exanthemata in infancy, particularly scarlet fever, measles, meningitis. The defect may be the result of deprivation in early life through disease or injury of two or more senses such as hearing and vision as the result of which the individual is robbed of the opportunity for contact with the outside world. Unless exceptional means are employed to communicate with him and develop his personality such an individual would remain defective.

Very little that is definite can be said concerning the pathologic anatomy of mental deficiency. The studies published by Fernald and Southard are perhaps the most reliable contributions to this subject.

Symptomatology.—A visit to an institution for the mentally defective gives one the best idea of the objective clinical manifestations of mental deficiency and I will ask the reader to imagine himself walking with me through the wards of such an institution. He will then observe in certain cottages little cots in which lie children of various ages in various degrees of crippled condition. The first thing that will probably attract one's attention is the unpleasant odor about such a cottage, for these children, although some of them are as old as six or seven or even older, have not developed those inhibitory controls which are necessary for cleanliness. They have an impulse to evacuate the bladder and bowels and they do it forthwith, without regard to time or place or the comfort of others.

The next impressions that will assail one will probably be those of strange noises, grunts, shouts, peculiar cries, and crude attempts at laughter. As one directs one's attention to any particular spot in the room from which a noise is coming one will notice that whatever the particular noise, it has a certain distressing and colorless monotony about it. It is expressed without any stimulation from without—merely as an inner impulse. The child lies helplessly with a blank facial expression, neither interested nor disturbed in any way by the noise which he emits. As one approaches more closely, one's attention will be attracted by all sorts of apparently meaningless and monotonous motions which some of these youngsters carry out. Some monotonously move a limb back and forth, others carry out some peculiar grimace, still others suck a thumb or other part of the body, and so forth. What will strike one as most unsual is that the faces of these children are so expressionless and indicate not the slightest vestige of any awareness of what is going on. If one should want to attract their attention one will probably succeed best by a loud sudden noise, for ordinary stimulation, even the stimulation of a pin prick may not do it. These children have developed very few neuron patterns and are in touch with the environment only in a very limited degree. These monotonous movements and actions are carried out by them for hours at a time; some of them seem to enjoy this monotonous activity because it is accompanied by a peculiar grunt of satisfaction.

Here and there one is attracted by a crude attempt at a word or two, usually badly pronounced and indistinct. One of course soon notices that the meaning of the word may be entirely foreign to the one who utters it. As one becomes a little more accustomed to the scene, finer details of the appearance of the children will attract one's attention. Some of them have large heads, others unusually small ones, the heads being wholly out of proportion to the body. Then again one notices some lying with limbs drawn up and fixed, others with limbs limp and lifeless, and only very rarely does one observe a face that might be termed attractive. Around the room older children will be walking about, with all sorts of peculiar mannerisms of gesture, gait and carriage. Their clothing will be in various stages of dishelvelment, and one notices ragged edges as though someone had chewed on them. Indeed one may be able to observe some of them in the act of chewing at their clothing or some other inedible objects.

This unlovely picture may be seen in any ward where the idiot children are kept, irrespective of the amount of care bestowed upon them. They are practically helpless, demand constant attention, are filthy and destructive and frequently succumb very early to some intercurrent disease. Even those neuron patterns which have to do with some of the most fundamental instincts, hunger and sex, do not develop to maturity in these children. Not only do the neuron patterns not develop but the potentiality for their development is lacking. The mothers of these children will tell you what difficulties they experienced in teaching the poor youngsters to take the breast, and some never learn even this vital act, or to hold on to a rattle, etc. From a social-economic point of view, as one can readily see, these idiots constitute the least problem. They are so helpless that they have to be taken care of from the first, usually in an institution, and by that very fact society is protected. Nevertheless, it does happen, now and then, especially where institutional provision is lacking, that idiots are the cause of serious disaster, particularly fire and loss of life. Some idiots attain physical maturity and vigor and with the mind of an infant may be the cause of much mischief.

As we proceed in our visit of the institution for the feebleminded, we come across small groups of boys and girls engaged in some form of simpler occupation about the institution under the supervision of some one, and are told that these children manage quite well at these simpler tasks, especially if they are of a restive nature. Some love their work once they have acquired the series of habits necessary for its carrying out, others have to be constantly urged on, while still others prefer to spend their days in idleness and mischief about the institution. We also note that a good many of them are neatly and carefully dressed, are able to conduct a conversation about simple objects quite interestingly, the majority being quite happy and contented. We discover, furthermore, that part of their time is spent in school or shop where they learn to read and write, even simple number work, and where they acquire a greater or less facility at some simple trade. In all of them, however, the thing that is lacking is the ability to plan and to form judgments. When confronted with new situations they are unable to utilize past experiences, past neuron patterns in new combinations which the adaptation to new situations demands. They lead what might be termed a habit life, naturally in some bad habits predominating, in others good ones, depending upon the nature of their early management and training. But habit, which James has called society's most precious conservative agent, and from the standpoint of the individual, "spinning out our fates, good or evil, and never to be outdone," is like fire, a good servant but a bad master. Habit is at once the strength and the weakness of the feebleminded. They form habits easily—good or bad—but it is very difficult for them to unlearn them. Moreover, unless carefully guided into habit formation they are apt to form what are known as impulsive habits, rather than those which are the result of deliberation, and impulsive habits are more often likely to be bad habits than good habits. Because they are lacking in reason and judgment and in the ability to deliberate and act rationally under novel situations, they are all creatures of habit. The wise and intelligent training of the feebleminded consists, therefore, largely in establishing in them good habits, and this means of course, not appealing to their reason, which they have not, but in requiring of them such action as will result in good habits. Left to themselves their habits are as apt to be bad as good, but under wise treatment they can be kept from forming bad habits and made to form useful ones. In this way their lack of judgment and inability to meet new situations is partially counteracted.

The same lack of judgment and lack of ability to adapt to new situations will naturally also prevent the feebleminded from modifying his habits to meet the demands of the situations. It is in this that one of the great values of institutional care for the feebleminded lies. In the institution they are not exposed to new situations, the environment is simple and largely unvarying, and the need does not arise for meeting new tasks. It is this also which makes the unsupervised life of the feebleminded in a busy and active community such a menace to others.

Other traits of make-up which are reflected in the behavior of the defective are weakness of will, lack of persistence in the accomplishment of a task, a wandering interest and difficulty in organizing one's energies to definite purposes. It will lead us too far afield to discuss here the nature of "will," but it might be said that the essential habits of attention and habits of action which are necessary for continuous application to a task lend themselves to develop but very meagerly in the mentally defective. The poverty of associations, the lack of clear-cut purposes, the inner distractibility and impulsive character of thought and action all contribute to this difficulty. Moreover especially in the imbecile classes, the impulsiveness of their reactions leads to an exhibition of an affectivity which brings these individuals in serious conflict with their surroundings. Suggestibility and imitativeness which we have already seen to be outstanding characteristics of child nature remain unmodified, or are manifested even to an exaggerated degree in the mentally defective.

In the lesser grades of defect, and as we approach the higher grades of feeblemindedness these defects are less pronounced, one deals essentially with the same phenomena. On the other hand, the less defective, because of the lack of an immediate necessity for institutional care, since many of them are not only able to care for themselves in a certain fashion but are not devoid of a certain degree of shrewdness which renders the condition rather deceptive, are apt to be a far greater menace to society. In fact, probably most of the serious social problems created by the defective relate to those cases which, without being definitely feebleminded, still suffer from a degree of dullness of intelligence which renders them incapable of forming the judgments and foresight, and from cultivating the inhibitory powers demanded of the individual in present-day life in a community.

The methods for testing the intelligence of the feebleminded are so well known that they require no description. The best statement of the situation is given by Terman.[11] But anyone who attempts to carry out an intelligence examination of a feebleminded individual will appreciate how important it is to note how he does a thing, what his attitude is towards a given test or toward the entire procedure, as well as noting what he can do. Such an examination is not apt to be an unalloyed pleasure, since much patience and tact is required in the face of the dullness and distractibility of these children and the difficulty they experience in rousing themselves to the required tasks. Repeatedly one discovers them staring unintelligently at the examiner, or looking vacantly into space, or restlessly moving back and forth in the chair. Questions may have to be repeated again and again and ordinarily the examination gets to be tedious both for patient and examiner.

One soon discovers the defective's difficulty in comprehending abstract concepts, his paucity of constructive imagination, that even in those who have acquired a certain degree of mental equipment it is not necessarily based upon anything more dependable than imitativeness and the result of reproduction, rather than the expression of native capacity for originality and initiative. Moreover, in contrast to the dynamic nature of the normal mind, the defective mind is static. That is to say, the defective has little ability to utilize such knowledge as he possesses for the purpose of acquiring further knowledge or intellectual growth, and fixed limits of development are commonly reached.

The two cases selected for illustrative purposes here reflect feebleminded individuals with especially active careers of anti-social tendencies. In one there is in addition the complication of sexual inversion. They are not to be taken as typical examples of careers of feebleminded individuals, since as we have already indicated, quite the reverse is apt to be true. But they do illustrate quite clearly what a combination of adverse social circumstances and unintelligent management is apt to do to the life of a defective.

Case I is that of a 24 year old Italian-American who was born in a squalid home, in a congested section of New York, of illiterate, simple-minded Italian immigrant parents, who had supported themselves by the combined earnings of the man as common laborer and of the woman as finisher of garments for a sweatshop. They led a primitive, almost vegetative existence, acquired only a few simple English words during their residence of thirty-odd years in the United States, and remained un-Americanized either in spirit or in fact. Of the 13 children which resulted from this union, four were miscarriages; a 27 year old daughter is profoundly feebleminded, still unable to tell time or make simple change and totally illiterate; a boy aged 25, formerly a member of the ungraded classes, is profoundly defective, is described by his school principal as "slightly unbalanced in mind, wild and uncontrollable," and has a Juvenile Court record. The subject of our present discussion, seventh in sequence, was born in New York on Aug. 29, 1893. The mother, though over-burdened with heavy domestic cares, was obliged to continue her share of industrial labor for the support of the home during her pregnancy with this child, as was the case also with her other pregnancies. But birth and early infancy appear to have been uneventful. General development appeared normal up to the age of five, when he was seized with whooping-cough, and later with a protracted attack of what appeared to be either scarlet fever or some form of meningitis. Recovery was slow and tedious and left him dull and markedly reduced mentally, necessitating a re-learning of walking and talking. On reaching eight, he had a severe attack of acute rheumatism, which invalided him for seven or eight months, and at the age of 12 he was confined to the Kings County Hospital with a severe protracted attack of typhoid fever, that still further reduced him physically and mentally. The effect of these severe inroads upon his health were reflected in his conduct at home, at school, and in the street. His mind at times seemed flighty, he was secretive, moody and uncommunicative. At school he was dull, inattentive, excessively truant, failed of promotion twice, and finally had to be sent to a truant school, and later to the Brooklyn Parental school. From early boyhood he came in contact with a totally unsupervised street life in a vicious neighborhood. Thus, to the life in a squalid, dark, unsanitary and crowded home, without bathing or toilet facilities in an old tenement building, there was added the influence of close contact with a neighborhood described by our investigator as follows: "Many cafes in the neighborhood frequented by bums who spoil boys. There is much drunkenness to be seen, many pickpockets, and not a few murders. In the neighborhood school across the street, 50 or more boys are on parole from the Children's Court." It is therefore not surprising that he early contributed to the police activities of the neighborhood. When not quite seven years of age, he was arrested in company with his brother for annoying neighbors. When not quite 10, was arrested for thieving but was discharged for lack of proper evidence; later in the same month was again arrested, this time for having thrown bricks at workmen in the neighborhood and for smashing windows, receiving a suspended sentence. At the age of 12 was arrested in company with several other boys for having driven off with a horse and wagon. The owner did not care to prosecute the case and the problem of the boy remained for the time being in abeyance. But it was only several months later that he was again arrested for having stolen some books from a book-store in company with some other boys, and was sent to the Catholic Protectory, where he remained about a year and a half. The high degree of suggestibility, which was later defined by laboratory methods, was reflected in his behavior by the ease with which he was led into mischief by the other boys of the neighborhood. No record of his behavior at the Protectory is available beyond the statement that he was in good health, could read and write, and was confirmed while there. At any rate, no significant change in conduct took place following his release, and seven and one-half months later he was committed to the House of Refuge for breaking into a fruit and grocery store with some other boys. He remained at this institution one and one-half years, when he was released after work had been found for him. His capacity to avoid conflict with the law did not improve as the result of his second institutional experience, and he was free only six months when he was sent to Elmira for burglary.

The monotonous repetition of the failure on the part of diverse social agencies, such as school and truant officials, police and court officials, and even officials of institutions of reformatory intent, where the boy was under observation for prolonged periods of time, to appreciate intelligently the serious social problem involved in this boy, suggests the desirability of a discussion of the subject of criminal responsibility from a new approach, wherein the culprit himself would play a relatively subordinate rôle. His contact with Elmira served for the first time in his career to bring out an effort at understanding on the part of society. He is described in the records of that institution as a coarse type of Italian-American, of good physical condition, poor mental capacity, poor susceptibility, poor appreciation, "a mentally defective pervert whose lack of parental control during formative period was undoubtedly the direct cause of downfall." An undersized neurotic with little or no working capacity, whose low moral status precludes the possibility of continuous effort. Properly environed, under inteliigent supervision, this juvenile delinquent might have become fairly self-supporting. The absence of incentive, however, coupled with a congenital defect, indicates a life of anti-social activities. He earned his parole in the usual time, maintaining average record. A really splendid effort at understanding this boy's problem, considering how the real issue had been ignored up to that time, but one wonders by what process of reasoning such totally opposed conclusions as having earned his parole, and indication of a life of anti-social activities could apply to the same individual. His subsequent life indicates that the predictions made at Elmira were justifiable and that, by the same token, the judgment that he had earned his parole was totally unwarranted. Is it possible that even the reformatory institution which is capable of rendering such dependable predictions has no choice of alternative beyond parole after an inmate has served the minimum required sentence? The boy immediately resumed his former mode of life upon his release from Elmira, was arrested on suspicion several months later, and about a year later was sent to the Penitentiary for about nine months for thieving. In serving his present sentence in State's Prison for grand larceny, he has reached the last in the round of reformatory and penal institutions which this state provides. That not a single one of these institutions had succeeded thus far in accomplishing what it professedly undertook to accomplish—the reformation of this defective boy—ought to raise the serious question as to whether any one of them was appropriately equipped to deal with the social problem presented by this boy, although they served well enough to satisfy the mere statutory requirements of temporary incarceration each time he came in conflict with the law.

His industrial career has been a total failure thus far, although in each reformatory institution an attempt was made to teach him a trade. It does not appear that he ever even attempted to work at any of these trades. His preference has always been for driving trucks and wagons, and when he was asked in the course of the examination what his ambition in life has been, he said, "I am crazy about automobiles." At one time he had accumulated $200, which he invested in a motorcycle; the boys persuaded him to exchange it for a motor-boat, and in the transaction he lost all his savings. In fact it never required much urging on the part of "the boys" to get him to do things. He realizes subjectively his incapacity to resist suggestions. "All the fellows have to say is 'Go on, Joe, don't be a coward,'" and he is ready to do anything.

We found him to be fairly well developed and well nourished physically, and aside from valvular disease of the heart, and deviated septum of the nose, free from disorders. Mentally he was very infantile, over-credulous and dependent in makeup. In reply to a request, he wrote the following letter:

"Dear Sir:

I am writing you this letter to let you know that I am feeling fine hoping you the same and in best of health. I was sent hair for Att. G. L. in the 2nd for 1 year and 11 months. I am going to be good and get out as soon as possible. I was send hear for Stealing a car, I would like to work wear I could learn more things about cars, because

I was always around cars. I will close my letter by doing so I send you my best regards and wish you in the best of health. From your truly,"

He coöperated well in the examination but was deliberately untruthful about certain matters and unable to give a correct account of others. Stream of talk was free, at times irrelevant and excessively circumstantial. Laughed childishly in relating his troubles, unconcerned about his incarceration beyond an intention "to be good and get out as soon as possible." Had very little insight into his twisted career and attributed his delinquencies to "the boys" who always made him do things. Never stole anything alone. Was afraid of the dark. People used to scare him with skeletons. Before he would steal he would have a peculiar feeling of nervousness which he attributed to fear rather than anything else. Parents were good to him, although mother was unusually severe. Would administer a severe beating on frequent occasions. Loved to travel in automobiles Was crazy about it, and when the boys suggested a joy ride, he stole the car.

The parents described him as mischievous, fond of children and animals, and was always bringing dogs home. Moving pictures claimed a lot of his attention. Laboratory examination showed him to be defective in general intelligence, memory very poor, learning ability very poor and learned chiefly by imitation, flow of free associations was retarded, and there was a certain degree of over-suggestibility. In a psychometric estimation attained a score of 67 per cent. out of a possible 100, an intelligence age of about 11.

It would be difficult, indeed impossible, to say what this defective boy's career might have been had his social surroundings and rearing been different. But the case illustrates clearly the point we have already made that to start with society had here a defective boy whose degree of social maladjustment is undoubtedly related to the manner in which his case has been dealt with by society. Fundamentally, of course, the fault was in trying to deal from a purely social point of view with what is primarily a medical problem.

Case II is that of a 19 year old negro boy, who experienced his first arrest at about the age of nine and who is at this writing serving his fifth sentence in reformatory and penal institutions, the last three for various sex offences. His ancestry is somewhat obscure. It is said that the paternal grandfather was a white man. The maternal grandmother is suffering from dementia following an apoplectic stroke. The father, who is said to have been a practicing physician, died of heart disease when inmate was four months old. The mother is described by our field investigator as approximately normal in intelligence, of slight education, suspicious, reticent, and unreliable in statements of fact. Attention shifting, extremely nervous, irritable, impulsive. In the past has had frequent attacks of nervous headache with occasional nausea, fainting spells, or momentary disorientation. The doctor called it "nervous prostration."

The boy is an only child, was unusually large at birth, alleged to have weighed 15½ lb. Birth and early infancy uneventful, except that mother alleges that when child was four months old, she fell with him and injured his head. He was bottle-fed until 14 months, and since early childhood has had free access to candy and other sweets. According to her statement at nine months of age several teeth were already carious from too much sweets, mother claiming that she had one of them extracted. The child, according to her story, was long in recovering from the nervous shock, became acutely nervous, irritable and fearful. On the approach of a man he would scream with fright, nearly going into convulsions. Mental development also appeared to have suffered retardation. When less than a year old (seven months?), while being spanked, went into a momentary convulsion, all the muscles growing suddenly rigid. This spasm has not recurred in later years. The boy usually lived in fairly respectable neighborhoods, but home conditions were very unfavorable, and from infancy grew up virtually as an orphan because the mother was obliged to earn her own and child's support by domestic service. Occasionally stayed with mother at place of service, but most of the time was boarded out with private family or with maternal grandmother, an invalid. The mother was always very stern with him while she had his custody; she consistently repressed him, allowing him no freedom of expression and keeping him very quiet and secluded at home. Until the age of seven, when placed in an orphan asylum, he had had practically no contact with other children. Mother stated that neighbors used to tell her that the boy was restrained too closely, and she now thinks they were right. She used various forms of discipline without discrimination. Frequently, in a fit of passion, she would use her fist on his head. In spite of her severity, he early became ungovernable, though corporal punishment was applied almost daily. Mother thinks that the effect of institutions, in which his later childhood was largely spent, has been altogether harmful, cowing him more completely and teaching him evil ways. He slept in the same bed with mother off and on until 11 years of age and until about the same age would wet his bed.

His recreational resources and activities were limited and confined almost entirely to moving pictures, of which he was very fond. He had no friends who called on him at home. His mother often urged him to seek the company of girls of his own age as other boys do, but he shunned them and knows no more of women than a babe does. He was long dependent upon others in dressing. In view of his later sex tendencies and experiences, which have already brought him to reformatory and penal institutions on three occasions, the behavior of this neurotic, ignorant mother, towards the inmate during his early life, is especially significant. Especially so the practice of sleeping in the same bed until the age of 11. She, however, blames her boy's downfall to his early contact with life in a reformatory institution, and asserts, "They learn all the dirty dirt they know in those institutions."

His schooling was very slight and extremely sporadic between 7 and 14. Though registered in several public schools during that time, he very rarely attended any, but preferred to spend his days roaming the street, frequently remaining away from home for days at a time, and refused at all times to study his lessons. In view of present findings of a pronouncedly retarded mental development, his failures were probably due largely to incapacity to carry the work demanded of him. He was in constant conflict with his teacher owing to his unruliness and misconduct in school. On at least one occasion he was expelled from school, in which connection the principal wrote the following letter: "P. is so disorderly that his teacher cannot keep him in the room. She sent him to me, and while I was out of the office, he opened my satchel, took my knife, 10 cents, and two pencils. It was only when I threatened to take off his clothes that he gave them up." Notwithstanding his mother's denial, his truancy appears to have been in association with others, who are characterized as "bad companions." Little can be ascertained concerning the character of his frequent disappearances, but evidently they greatly worried his mother, who repeatedly secured his commitment to an institution as ungovernable, disorderly, etc.

His first contact with the law occurred before he was nine years of age, when complaint was made that he was a disorderly child. Disposition was deferred. Several months later was sent to the Brooklyn Training School on a similar complaint. After about six months was discharged upon the request of his mother. His behavior did not improve and in December, 1909, was again committed to the Brooklyn Disciplinary Training School. It would be of interest to know what if any effort was made during either of his sojourns at the disciplinary school to ascertain the underlying whys and wherefores of his incorrigibility. In February, 1911, his mother again made application for his discharge, whereupon the superintendent of that institution wrote to the Society for the Prevention of Cruelty to Children as follows: "I wish to say that this boy has a satisfactory record, has been here a sufficient length of time, and I am satisfied to recommend him to the Committee on Discharge for parole." The boy was accordingly discharged.

It would, clearly, be out of the question to have expected the boy's ignorant and neurotic mother to appreciate unaided the causes of her boy's difficulty. One might, perhaps, be inclined to view charitably the naive way in which the over-burdened teachers handled his problem, that is, by expelling him from school; but one is constrained to question the good judgment exercised by the Parental School authorities in discharging him merely because he had "a satisfactory record and had been there a sufficient length of time." Yet the action of this superintendent was only typical of the action of other authorities who handled this feebleminded boy's case subsequently. The superintendent of the reformatory where the boy had been sent for having committed an indecent assault upon a three year old girl child, wrote to the above-mentioned society that the boy was to become eligible for parole on Oct. 24, 1914, and wanted to know whether there were any objections to his release. In reply the society wrote: "We would suggest that before he is paroled it might be well to have him examined by your physician who could determine whether or not he would be likely to commit any further acts of a perverted nature when released. If in the opinion of the physician it is safe to permit the boy at large, and if his record at your institution is such as to be favorable for his parole, the society would offer no objections to the boy's release." It might have been difficult or impossible to predict with certainty at that time whether or not the boy would ever commit a similar act, but an intimate study of the boy would have brought to light at least the fact of his feeblemindedness, and a prediction based on such information would have indicated continued supervision if not actual custodial care. As a matter of fact, the boy was discharged. When less than six months later he was again arrested for a particularly vicious sex assault upon a boy aged eight, and sentenced to 20 years to the New York State Reformatory at Elmira, his problem was not brought substantially nearer solution. Although his crime apparently provoked ample indignation in the judge who sentenced him, considering the "punishment" imposed, the institutional authorities in their turn saw fit to release him after 14 months because, forsooth, he had earned his parole. This proceeding on the part of the Elmira authorities is the more inexplicable as, in contrast to the agencies which had previously handled the boy more or less blindly, they had means of knowing precisely what his future career was likely to be. To quote from their own records: "A coarse type of mulatto, mental status poor; capacity poor; stability poor; moral appreciation poor; susceptibility to training poor." And this is what they had to say concerning his future: "Prognosis: A menace to society; permanent custodial care indicated." Why then did they release him? To be sure he doesn't belong in a reformatory—he belongs rather in an institution for the feebleminded; but should not an attempt have been made to have him committed to such an institution in accordance with the doctor's recommendation, rather than to ignore the facts and release him forthwith into the community? Less than four months after his discharge from Elmira, he was again convicted of a perverse sex assault upon a boy aged 11. At the time of his arrest he admitted to the police that he had used four other boys in a similar fashion. This time he evoked the wrath of the judge, who, accepting a plea of sodomy as a first offence, sentenced him to 10 years and 6 months in State's Prison, this notwithstanding the boy's known delinquent record.

His industrial career has been extremely inefficient and on a par with his low capabilities. His average length of service in a place was three or four weeks.

We found the boy to be well developed and free from physical disorder. Mentally he was very defective and inaccessible. He comprehended the least complicated questions with considerable difficulty, attention was difficult to gain and more difficult to hold for any length of time. Memory was very much confused and unreliable, and affected to a pronounced degree by suggestions from the examiner. Seemed to be considerably taken up with internal distracting thoughts. In the midst of a conversation during which active attention on his part was demanded, he would become preoccupied, lose himself in a sort of dazed state, and when addressed would pull himself together with a start. Orientation for time was uncertain. Emotional state was extremely dull and indifferent. Did not seem to be affected in the least by a discussion of his past criminal career, nor with intimate probing into his sex life. His insight into his difficulties was meager and limited by his restricted intelligence. Claimed to have had first heterosexual experience at the age of 10 with a girl in the neighborhood, but has always felt a distinct preference for the male sexual object. Insists that he had his first homosexual experience at one of the reformatory institutions, where, he says, this practice was very prevalent among the boys. Has made efforts to abstain from this practice but was incapable of doing so. He would get a feeling all of a sudden that he wanted to do it, would be uncomfortable, restless, and could not sleep until he did it, when he would experience a marked sense of relief. This urge apparently was of great potency, since at times he would stop at nothing to gratify it, even threatening to murder the unwilling victim of his assault. He had no conception as to what his future behavior in this respect would be, nor any interest in shaping his future. A pyschometric examination gave him a mental age of 11.5 years.

More recent studies of the mentality of reformatory and prison inmates has led to much discussion concerning the precise extent of feeblemindedness among these individuals. The above two cases ought to indicate quite clearly that this aspect of the situation is only of secondary importance after all. What is important is to recognize the great mischief which is involved in failing to deal with such cases, who at the bottom have a medical issue as a determining factor, medically, with results of constructive adjustment rather than what is reflected by the above cases.

This brings us to the question of treatment. It must be clear that this involves both individual attention to the defective and the social management of the problem of feeblemindedness.

With reference to both aspects there is a tremendous literature available and it is really beyond the point here to go into details.

The treatment of the individual case always involves an educational approach and efforts at appropriate social adjustment. The one has for its aim the bringing out in the individual to the limit such capacities as he possesses with attention as far as possible to give practicability to this training. Thus, much more so than is the current practice in the education of the normal the teaching of the defective should have for its aim its availability for adaptation to the tasks of life. The outstanding issue in the social adjustment is to fit the demands put upon the defective to his capacity for meeting them. Frequently enough this necessitates a type of modification of these demands which can only be had in a custodial institution.

But in many cases approaches of a purely medical nature are indicated. Especially imperative does it become nowadays in view of the pre-occupation with the subject of endocrinology to search to the limit for a possible defect along these lines with a view toward its correction. Although our knowledge along these lines is still extremely limited, evidence is accumulating which justifies a good deal of experimental empiricism along endocrinologic lines. Whenever the physical and metabolic findings suggest at all an imbalance in this direction, especially where no definite other causes are discoverable for the mental defect, recourse to endocrinologic treatment is thoroughly justified.

As regards the broader, social management of the problem the achievements of the State of Massachusetts deserve special attention. These are clearly outlined in the paper by Fernald already referred to.

THE CONSTITUTIONALLY PSYCHOPATHIC CHILD

A comprehensive discussion of this rich field of constitutionally burdened children should involve a critical re-estimation of the views which led to the acceptance of a constitutional basis for these anomalies of personality makeup. The literature on the subject is pregnant with rich clinical descriptions and much evidence to bear out the contention that one is not dealing here with a disease process *per se*, which has its onset, progression and termination, but with an anomaly of personal makeup sufficiently different from what one ordinarily meets with in normal individuals to justify viewing it as abnormal. This peculiarity of makeup comes to light already during early childhood and tends to persist without significant modification throughout life. The consensus of opinion among the older students, at any rate, is to the effect that only very rarely, if ever, does one encounter here a definite trend towards improvement or cure, or any tendency to progression in the nature of a deterioration such as one is familiar with in some of the psychoses. Such deterioration as one may observe in the course of a psychopathic life is rather in the nature of a social-economic deterioration which is inevitable in the case of those psychopathic individuals who are not very favorably situated economically.

Nevertheless, recent researches in the field of metabolism, especially as regards the place in the individual's economy of the glands of internal secretion, and the greater recognition which is being accorded the ordinary conditioning influences of a psychological nature are challenging very seriously our conceptions concerning psychopathy. The belief is growing that many of the pathologic traits of the psychopath which we were wont to consider as expressions of innate endowment may have been induced by adverse conditions in the life of the child and that the fundamental instability of organization of the personality of the psychopath may be related definitely to some endocrinologic imbalance.

For our present purposes it would be beside the issue to enter into a more detailed discussion of the merits of these contentions beyond saying that they justify a thoroughly experimental attitude in the matter of treating these conditions, since the element of immodifiability is not to be taken as rigidly as formerly.

Definition.—The term "psychopathy" is sometimes used in a general way to designate various forms of mental aberration. As used here it is synonymous with such terms as "constitutional psychopathic states," "psychic constitutional inferiority," etc., and refers to a chronic state of mental and social inadequacy leading to chronic, abnormal social and mental reactions which cannot be classfied under any of the known categories of mental disease or mental defectiveness. While occasionally a constitutionally psychopathic individual may also manifest some defect in intelligence, the majority do not show such defect and one encounters amongst them now and then individuals of superior intelligence.

A practical distinction between the constitutional psychopath and the mentally defective is to be found in the fact that whereas the former, though ordinarily socially inadequate and sometimes definitely antisocial in his tendencies, may rise to exceptional heights of achievement, the latter can never become an exceptionally valuable citizen. History records the lives and achievements of many a noted man who undoubtedly displayed all the earmarks of a constitutionally psychopathic individual. As regards the relation of this condition to mental disease proper this might be said: While the condition itself cannot be considered in the light of a mental disease, acute reactions of the latter type are relatively easily brought out in the psychopath by undue environmental pressure. Thus under confinement in prisons or military camps, especially when a rigid discipline is maintained, the psychopath may develop an acute mental disorder of a profound but transitory nature.

The distinguishing feature of the constitutionally psychopathic state lies in a defective organization of the instinctive-emotional life, in an anomalous organization of the personality in the nature of a fundamental disharmony affecting primarily the emotions and sentiments, the imagination and the will. In addition to the inadequacies of a psychologic nature, there are readily discerned in a large number of cases significant disturbances of a physical and physiologic nature. One observes frequently a general restlessness and lack of physical poise, poor muscular coördination with a strong tendency to queer habit movements, poor general development, flabby, atonic musculature, irregular and disproportionate development of parts of the body. Physiologically, one encounters irregularities in the onset of puberty and in the menstrual functions, more commonly a precocious onset, periodic headaches and attacks of dizziness, delayed development of sphincter control, defective organs of special sense, etc.

It is particularly these manifestations which are suggestive of a closer relationship than is ordinarily recognized between these conditions and the more clear-cut endocrinopathies.

Clinical Manifestations.—Attempts to characterize the psychic makeup of these individuals have been many, but it is still difficult to supply a well ordered statement of the situation.

First of all one meets with the marked irregularity and disharmony in the development of the psychic functions, with significant variations in total adaptive capacity from day to day. Side by side with good, and sometimes, even superior intelligence, there exists a degree of emotional instability and weakness of will which precludes any possibility of adequate social adaptation. Moreover, as a result of slight changes in environmental conditions, and at times, even under stable environmental stiuations, the psychopath shows a significant variation in capacity for performance from what is usual with him, which cannot be accounted for on any other basis except on the basis of a profound disturbance of his internal economy. This leads us to other outstanding traits in the makeup of the psychopath, namely, his great impressionability and suggestibility to external influences. One is inclined to attribute this, not so much to lack of intelligence and inability to form independent judgments, but rather to a peculiarly sensitized state of the organism which leads to a high degree of innate responsiveness to external impressions. It is perhaps part of this same quality in their nature that they tire so readily of a more or less commonplace routine of daily life and show a marked attraction for new and unusual experiences. Hence their tendency to vagrancy, nomadism, criminal exploits, especially those involving personal danger, addictions of various sorts, sex experimentalism. They particularly chafe under confinement, where the daily routine is more or less monotonous, and are apt to exhibit under these conditions marked affective outbursts.

Taking the life-career of the psychopath as a whole, it reflects a marked inconsistency and inferiority of "character." Their conduct does not seem to be governed, to any great extent, at any rate, by well-established inner directives based upon well-organized attitudes and sentiments. They tend to exhibit a pathologic degree of levity and frivolity in their attitude towards life, a peculiar, childish inability to guage the seriousness of a situation.

They are incapable of pursuing consistently a line of action for any length of time, so that it is difficult to discern in their life-performance any definite goal or objective. They frequently show significant discrepanices between stimulus and reaction. The behavior reactions of the psychopath when viewed objectively appear to the observer as queer, odd and out of place, as though a raw, sensitive surface had been suddenly irritated, leading to intense and passionate reactions. Equally common is the tendency to "over-valuation" of more or less ordinary events. They show brief but intense enthusiasms. What ordinarily provokes interest becomes with them a passion, an appetite readily turns into a lust. The psychopath has an extreme attraction for those pursuits which tend to bring out intense states of being "stirred-up," such as gambling and other speculative enterprises of a hazardous nature. They are subject to pathologic fluctuations of mood, which seem to be determined more often by inner causes than by environmental changes. These mood fluctuations are very pronounced, sometimes exhibiting a true cyclothymic makeup. They have a strong penchant for the fantastic and unreal. It is my personal opinion that the psychopath's strong inclination for the "evasion," the "lie," the "subterfuge," is to be looked upon as a genuine fear of, or timidity in the face of reality, a condition which is not uncommon in the reactions of specially intimidated children. These individuals are apt to be rather intolerant, suspicious, querulous and contentious, readily falling into a pseudo-delusional and delusional interpretation of events. Finally, one must speak of the unbounded egotism of the psychopath, a personality trait which tinges all his reactions.

The above character traits of the psychopathic personality are naturally much more clearly exhibited in the adult than in the child. Or one might put it more accurately, perhaps, that they impress us with a greater abnormality when we see them displayed by the adult, since much of that which we recognize as abnormal in the adult psychopath might be looked upon merely as an infantilism and immaturity of his makeup. He is essentially an immature and unsocialized egotist whose egotism might be looked upon as a compensatory endeavor for a real or imagined inferiority. But although these manifestations might be considered in a large measure as mere exaggerations of normal traits and as characteristic in many respects of the childhood period, we do encounter in children personality makeups which are more or less identical with the makeup of the adult psychopath, and one is justified in speaking of them as constitutionally psychopathic children.

Classification.—Largely perhaps out of a desire to bring some order into the many-sidedness of the clinical manifestations of constitutional psychopathy, various attempts have been made to classify these individuals into types. The most noteworthy of these attempts was made by Sholz[12] in his book, "Anomale Kinder." He divides them into the indolent, the depressed, the excited, the periodic, the emotional, the impulsive, the unstable, the eccentric or perverse, the fabricators and liars, the obsessional, the morally inferior, the sexually abnormal. No matter how openmindedly one might approach Sholz's otherwise very excellent presentation of the subject, one cannot escape the feeling that his classification is altogether too artificial and strained. Clinical experience is more in accord with viewing the psychopath as an individual capable of exhibiting the characteristics of all these various types. Herman Adler[13] classifies the psychopath into the paranoid, the inadequate and the emotionally unstable personalities. Most of the traits of these individuals can be grouped under these categories, and there is some justification for the attempts of students to classify these abnormal individuals.

The point which is not receiving sufficient consideration in the available literature is to what extent these psychopathic traits may be attributed to faulty developmental experiences. The assumption that the condition is innate and hereditary is justified in many instances, but even when one is dealing with such cases there frequently emerge significant social-psychologic elements in the early life histories of these individuals to justify the belief that some, if not most of their abnormal traits, are definitely conditioned by life experiences.

Thus, in the case history that follows, one can readily discern significant issues of a hereditary nature, important assaults upon the personality through repeated illnesses in childhood, certain anomalies in the structural makeup of the child, all of which substantiates the belief that one is dealing here with a constitutionally inferior child. But there are also present in the case very important psychologic issues which must be taken cognizance of in the total estimation of the personality of the child:

A boy of nine of Russian-Jewish stock was referred on Feb. 16, 1922, for examination by his boarding mother because of his lack of coöperation at home and lack of responsiveness to her efforts to guide him. He gets cranky and excitable very easily, is not frank, prefers to crawl away into a corner without saying much to anyone. He does not mix well at play with the neighborhood children and is particularly abusive to his older brother. She feels that the boy is decidedly abnormal, though according to her statements, he is very bright at school and very conscientious about the home work.

The boy is very badly burdened hereditarily. The mother, whom the father describes as irritable, dissatisfied and always worrying about one thing or another, died in June, 1921, in a private sanitorium for mental diseases following scalds received by accidentally falling into a tub of hot water. Nothing definite is known about the nature of her disorder, but according to the boarding mother, she had been insane long before her commitment to the hospital, and when our patient was five, she sent both boys to a boarding school in Pennsylvania, saying she was unable to care for them. She was so unmanageable that she had to be tied down to her bed at the time. An attempt to diagnose her mental disorder must take into account the fact that she had been called diabetic because of a 5 per cent. sugar content in the urine, had had a positive blood Wassermann reaction and was in the midst of her menopause. From the description of her disorder, it would appear to have been related to the manic-depressive group. A maternal uncle suicided at the age of 20, the cause of the death of another is unknown. A maternal aunt died of cancer. The maternal grandmother is a coarse, suspicious woman of 70. Nothing significant on the paternal side except that the father is a coarse, rather nervous type of individual, who at one time had a positive blood Wassermann. The patient's brother, about one year eight months his senior, did not talk until three or four years of age and the family thought he never would walk. A year after entering school he became "like paralyzed," could neither stand nor do anything. He began to improve after several months. According to the father, the doctors did not diagnose the case. This boy is quite well now, attends public school and is in 5B.

Our patient's birth was uneventful. He was partly breast and partly bottle fed, walking and talking markedly delayed. The delay in walking was attributed to flat feet. When quite young he had measles and when about seven was diagnosed choreic. Wassermann blood test since the mother's death negative. Nothing very definite is known of the home background prior to the age of five, when he was sent to boarding school, except that conjugal relations of the parents were not very cordial. The mother apparently had a poor opinion of her husband and regretted her choice immediately after marriage. She constantly criticized everything he did, worried a lot over the children and maintained that he did not care for them at all, though at times he did things she disliked, to spite her. He characterizes their conjugal life as very unpleasant.

The character of the boarding school the children went to is not known, but according to the father, our patient has been afraid and timid ever since. Following the mother's death, the father intended to place the children in an orphanage, but was dissuaded from doing so and instead, placed them with the present boarding mother, in a clean, well-cared for apartment, with standards of living considerably above those the children had been used to. The family in the boarding home consisted of the mother and two children, a boy of 13 and a girl of nine, and the mother's mother. The boy has frequently complained to his father of abuses by the boarding mother's children, who call him fool and crazy and of the lack of understanding on the part of the boarding mother. He cites as an illustration the fact that because of some difficulty with the boy, she has not spoken to him since the last week in February. He cautiously expresses the feeling that the children are neglected, though not intentionally so, but because the boarding mother expects too much of them, forgetting that they are only babies. It is difficult to estimate accurately the dependability of the father's opinion, though during the last interview with the boy on March 6, he appeared to be very unhappy and miserable, and the father attributed his unhappiness to dissatisfaction with the boarding home, adding that the boy visits him practically daily and always expresses the hope that he might come home to live with him. The boarding mother on the other hand is quite evidently a more refined and intelligent woman and although not free from a rather evident dislike for the boy, persuaded the father of her own accord to have the boy examined and is apparently anxious to have the boy receive medical care in the hope of improving his personality and behavior. She appears to be genuinely concerned about the boy's welfare though obviously "fed up" on what she calls his mean disposition. She states that she had a great deal of trouble from both boys when they first came to her because they were like animals without training or manners. They ate soup with their hands, dipping in their fingers to pick out the vegetables which they stuffed into their mouths until they could hold no more. When he thinks no one sees him, the patient still resorts to these methods.

Thus the patient's home background might be summed up as unintelligently neglectful because of disharmony and low standards of the parents and probable psychopathic makeup of the mother until he was five, then a period in a boarding school, which according to the father left him timid and afraid, then a period of neglect and anxiety during the mother's active illness and finally his present environment in which he is unhappy and unresponsive, not at all unlikely, because of the sudden transition into a setting of much higher standards with consequent demands for making new and difficult adjustments.

The parental home is located in a colored neighborhood which the father describes as thoroughly bad, but the boarding home is located in a better neighborhood.

Very little is known as to his early companionship and recreational outlets, but this has probably played no significant rôle because of the boy's makeup. It is not known definitely when he began attending school, but according to his father, he entered kindergarten when very young, has made normal progress and is at present in 4B. His teacher has considerable insight into his makeup and is very tolerant about some of his behavior tendencies. The boy bears the reputation of always having been studious and anxious about his lessons. He has B+ in every subject and will probably improve this to A. He is especially good in arithmetic. The boy has expressed the hope of graduating from school with highest honors and of beating his brother by getting through a year sooner. According to the boarding mother, the father encourages this attitude in the boy.

Our examination shows a well-nourished, pink and white complexioned boy of nine, with an unusually large head. His wing-like ears, broad mouth and jaw, give a certain oddness to his appearance, which is aggravated by a left external strabismus. His teeth are irregularly placed, upper canines poorly formed and simulating lateral incisors. Central incisors broad and notched. There is a postural lordosis and kyphosis, with flaring scapulæ and broadening of ribs, left shoulder lower than right. Cervical and inguinal glands are palpable, as well as right epitrochlear. Genitals are small, with scrotal fold. The chest expansion is fair, lungs and heart normal, blood pressure 95/80. There is some hesitancy in formulating words. He exhibits a thymic-pituitary endocrine state. He was rather slow and awkward in dressing and undressing and inclined to dawdle very much.

During our first interview with him on Feb. 9, 1922, he was rather shy and infantile in his attitude, but soon began to respond better and became friendly and thoroughly coöperative. During the interview he was very fidgety and restless, and every once in a while would appear to be lost in a day-dream, staring vacantly into space. Attention, however, was easily gained, he showed no difficulty of comprehension, and at times exhibited quite a healthy sense of humor. His attitude towards his mother's mode of death was rather curious in that in conversation about this, he emphasized mostly the fact that his father was suing the sanatorium for damages. He realized that he was a little nervous at times because he could not sit still, but said he was not worried about it. His attitude towards his present home during this interview was decidedly in contrast to what his father said it was.

He stated he was happy where he was, that he was particularly fond of the boy of the family, and that he was having a very nice time of it. Sometimes he does not like to play with others as he prefers to go off by himself and read. He is very fond of reading and emphasized how conscientious he was about his home work. The father, however, stated that he had noticed that the boy was afraid of telling his true attitude towards the boarding home as he fears it might displease the boarding mother and it would not do him any good.

A better idea of the boy's personality makeup can be had from descriptions of those who observe him in his daily reactions at home and school. His teacher has noticed that he was "nervous and peculiar in ways." For instance, all of a sudden, he is apt to get out of his seat and sit down on the floor. Sometimes during the lesson, he turns around and looks out of the window. Usually he is by himself and keeps away from all the other children. Otherwise he is very good, conscientious about his work and "all attention" in class. According to his father, the patient never played much with other children, was always quiet and by himself. If left alone he would make no effort to satisfy his wants—if not offered food, he would go without eating and not ask for it. His immediate response is generally antagonistic, but sometimes he relents afterwards. He is selfish about his belongings and unreasonable in his demands regarding those of others. He cannot play with other boys because he is always right and they are wrong.

His boarding mother describes him as sullen, contrary, antagonistic and rude; and once, when angry, he kicked his father. He responds better to sternness than gentleness. He gives a great deal of trouble in connection with food, some things he does not eat because they have skin, etc. He feels he is unjustly treated and criticized even when this is not the case. He instigates all kinds of mischief, is clever about covering his own tracks and then stands calmly by while others are blamed for his misdeeds. He does not fall asleep readily and is awakened by the slightest noise. He likes to help in the house and when thus occupied, seems to be more cheerful and talks at considerable length.

A psychologic examination given Sept. 26, 1921, at the Neurological Institute gave him an Intelligence Quotient of 99. He was noted to be exceedingly self conscious and somewhat stubborn, but it was noted in addition that mentally the child does not appear to be in any way unnatural except for the fact that his range of knowledge was narrow.

Some indication as regards predictability in this case was gained in connection with our efforts to help the boy to a better adjustment. At no time did we feel that we had succeeded in achieving a thoroughly workable rapport with the boy. In the midst of a quite pleasant and interesting conversation, he would sink into a sulky and irritable mood, and would either absolutely refuse to answer questions, or show a strong tendency to an active negativism by doing just the opposite of what one asked him to do. In connection with these changes in mood, the facial contour and coloring would change and much objective evidence was gained of the deep emotional disturbance which must have gone on inside of him.

The father repeatedly complained about what he called the boy's "mulishness," which would come on, no one knew whence or why, and render any intercourse with him absolutely impossible. He would get on all right as long as the path of daily adaptation was kept smooth for him, but as soon as any obstacles came his way, his intense egotism made adjustment very difficult or impossible.

In spite of these indications of a rather bad prognosis in this heavily burdened, constitutionally inferior youngster, it would be decidedly worthwhile to pursue the endocrinologic treatment which had been instituted, and to endeavor to provide for the boy a kind of home setting which would exert an intelligent and sympathetic, though consistent daily discipline directed to a greater socialization of this boy's reactive tendencies.

The next case is that of an exceptionally bright boy of nine and a half years of age, of Russian-Jewish extraction and native birth, who was referred by the Jewish Board of Guardians because of his persistent fighting with the other children in the block, his disobedience and disrespectful attitude towards his parents, his persistent stealing, bad behavior at school and general nervousness. Various forms of control, including physical punishment, had brought no results.

The boy, an only child, is the issue of rather nervous stock on the mother's side. The maternal grandfather was a nervous, irritable man, subject to "fits of depression." The boy's mother, a fairly intelligent woman of 37 is restless and explosive in manner, talks loudly and rapidly, and is easily excited and irritated. She is a very much disappointed woman, because of her husband's colorless personality and failure as a business man, is extremely over-solicitous where it concerns the boy's problem and finds her chief emotional outlet in worrying and fussing over him.

On the paternal side there is a defective cousin who required a bond before being admitted to the United States. The father is a quiet, colorless person, a complete failure in business, totally dominated by his wife and lately, in connection with an accentuation of their troubles, has found refuge in a gastric neurosis which keeps him incapacitated for much of the time. He has also shown a progressive decline in mental capacity and has become practically unreliable, even in small business matters.

The couple were married about 15 years ago, about a year after their arrival in this country, and our patient was born six years after their marriage. Pregnancy and the boy's birth were uneventful, he weighed 7 lb. at birth, was both bottle and breast fed, began teething at eight months, walked at 15 months, and although he was saying single words at 18 months, he did not form sentences until three years of age. For the first seven or eight months of his life he was a very restless baby, crying a great deal for no apparent cause.

He had none of the children's diseases until he was seven years of age when he had measles, with which he was ill for four weeks. Soon after recovering he had an altercation with a boy considerably older than he. The boy's mother interfered, striking our patient on the head, whereupon the patient's mother rushed out and struck the boy down. This lad, who was said to have had a weak heart, died two hours after the incident. Characteristic of the makeup of our patient's mother, is her considerable preoccupation afterwards, with ideas about suing the other woman for damages.

The mother dates the boy's difficulties from the time of this incident. She stated that that night he woke with a terrible cry and had what she terms "hysterics." For four days following he had similar spells. His eyes were red and swollen, and during the night he cried a great deal and complained that he was afraid of ghosts and burglars. Aside from a light case of scarlet fever shortly before his contact with us, he has had no other illnesses.

For the past five or six years the lad's environment has been very bad from a physical as well as a mental standpoint. During this time they have lived back of their little music store, in three small, overcrowded, dark and very depressing rooms. The boy is a constant witness to the bickerings and discussions of domestic affairs between his parents and to the constant complaints of his mother about "all she has had to stand for during the past 14 years of their married life." The home lacks even the slightest kind of harmony and peace, and provides no privacy for anyone. The father, mother and our patient sleep in one room. They eke out their living in running a little music store in which business the wife plays the dominant rôle, as she does in everything else, since she does not trust her husband's ability and is in constant fear that he may short-change himself.

The parent's attitude toward the boy's problem has been difficult to define accurately. They are both very much concerned about his misconduct, deeply hurt by the disgrace which he brings upon them and quite vociferous in their intention to do everything possible for him. But when it comes to coöperating in carrying out even the simplest suggestion in connection with the boy's readjustment, both find themselves very much hampered by the emotionalism which they bring into the situation.

The boy's chief behavior difficulties, as has already been indicated, have been his stealing and fighting propensities, extreme restlessness and total lack of response to corrective measures. He became hard to manage shortly after the blow on the head. When corrected, he stamps his feet, screams, but when threats of a whipping are brought into the situation, he cries and begs for mercy and promises to change. But these professions of good intentions never have materialized in anything more constructive. About a year before his contact with us, be began taking money from the till in the store. From that time he has stolen anything he could lay his hands on, spending the money thus obtained on mechanical toys, candies, and as will be seen presently, on more ambitious ventures.

Some time before he came to us his attention was attracted by an advertisement of a miniature moving picture machine, which was to be had for $10, and to obtain the machine, the lad forged his mother's signature to a check for this amount. Later he forged another check which he sent off for some "health and strength" giving medicine which he saw advertised in the papers.

Our little patient has always been clean and neat in his habits, only on rare occasions wetting his bed, after having been reprimanded. Though he was kept in dresses until four, and his mother tried in various other ways to make a little girl of him, he showed no evidence of any sex pre-occupations or interests. In this connection it is of considerable interest to note that as the mother finally got herself to follow our suggestions for a consistent elimination from the boy's daily regime of the definitely hampering affective indulgences of the boy, he developed the tendency to crawl into her bed at night, became fearful of going to the toilet by himself, insisting that she accompany him. Our difficulty in getting this disappointed and emotionally starved woman to see the real danger of a too intense affective relationship between the two, constituted one of the main stumbling blocks in managing this boy's problem.

Our contact with the boy revealed a frank, very talkative, and sociable lad, extremely sophisticated and glib in his discussion of current events as gleaned from the newspapers. He was of a decidedly manic temperament, restless and active, with considerable pressure of speech. His mental content was made up largely of matters considerably beyond his age, and to a large extent with the relative merits of various strength and health giving medicines which he has seen advertised in the newspapers. He would like to be strong and rich, and speaks of rather elaborate altruistic plans when he achieves these goals. The boy is also very much pre-occupied with exciting scenes from the movies which he attends frequently, in fact whenever he has the price. According to his story he makes friends easily and is popular, but shows intense envy of the economically better favored children with whom he comes in contact at school.

Physically there was nothing especially outstanding in his make-up except a rather marked visual defect in both eyes, and from an x-ray examination of the skull, doubtful indications of an increased intracranial pressure were present, especially in the frontal regions.

The psychologic examination of the boy showed him to be very superior intellectually, with an intelligence quotient of 118. He was very spontaneous, coöperative and showed a remarkable degree of auto-criticism for a boy of his age. He comprehended fully the social implications of his conduct, but somehow could not exercise the necessary control and inhibitions for better behavior.

In this case, even more so than in the preceding one, it is difficult to determine how much of the boy's difficulties are based upon innate and unchangeable characteristics, and how much the decidedly deleterious, environmental stimuli have to do with his difficulties of adaptation. The head injury, of course, must be taken into consideration, since we know that these injuries, even when mild, may be followed by changes in character and behavior which justify us in speaking of a post-traumatic psychopathic state.

The problem, on the whole, seems to be that of an extremely active, mild manic temperament, unfortunately placed in an environment in which he is constantly over-stimulated, both by general domestic affairs, and the nagging of an over-solicitous, over-excited, irritable and disappointed mother who finds her chief outlet in worrying and fussing over the boy. The situation is further aggravated by the father's rather hopeless attitude toward life in general. Some progress has been achieved in modifying the boy's attitude toward his stealing propensities, and he has abstained recently from these practices, but his general reckless behavior, pugnacity and nervousness remain quite unchanged and are continually conditioned by his irritable mother.

THE PSYCHOSES OF CHILDHOOD

As has already been indicated earlier in this chapter, frank mental diseases are relatively uncommon in childhood. When they do occur they are characterized in the main by the same symptoms and by the same tendencies as regards course and termination as in the adult. Because of the relative infrequency of these manifestations and the necessary limitations of this chapter, our discussion of this subject will be brief. It is necessary, however, to give a brief survey, at least, of the nature and meaning of the more common manifestations that go with the frank psychoses. Those who are unfamiliar with the essentials of normal psychology, a prerequisite for a proper understanding of the abnormal manifestation of mental functioning are referred to Dr. Goddard's[14] book on the subject for an exceptionally clear and concise statement of the principles of normal psychology.

1. DISORDERS OF CONSCIOUSNESS

Consciousness may be impaired or weakened, and we speak of it as clouding of consciousness. It may also be entirely abolished—unconsciousness; or exaggerated—hyperconsciousness. Unconsciousness is a phenomenon of dreamless sleep, of a comatose state, such as following severe head injury or apoplexy, or of profound stupor as in intense drunkenness. Clouding of consciousness is a manifestation of many mental disorders and has as its chief accompanying symptom more or less complete disorientation. When we are normally oriented we possess a clear notion of our own personality. We know who we are, we know certain vital facts of our own lives, etc. We furthermore have a clear notion of the outside world, and of time. Now a person may be disoriented in any or all of these three spheres and we speak of him as being disoriented for person, or for place or for time. In delirium tremens, for instance, the orientation for time and place is lost, while that for the person remains intact. It is ordinarily quite easy to detect complete loss of consciousness. The person is absolutely indifferent to his environment, "dead to the world," so to speak, and does not respond even to the strongest stimulation. However, the condition known as "catatonia" resembles unconsciousness very much, but is not so in reality, as the patients in spite of the appearance of being dead to the world are more or less fully aware of everything that is going on about them. Mere clouding of consciousness is detected through asking the patient certain questions intended to elicit information concerning his state of orientation.

Disorders of consciousness affect conduct very materially. Complete loss leads of course to absolute inaction, lesser degrees of clouding of consciousness may impart to the actions of the patient a more or less impulsive character. The brutal and ferocious behavior of some epileptics while in a semiconscious state is due to this. Clouding of consciousness unless accompanied by marked excitement and disorders of perception renders the individual emotionally indifferent and dull—and reduces the will to action. When accompanied by hallucinations and illusions, as it frequently is, or by delusional states, or by fear and anxiety, the emotional state and behavior will be in accord with these symptoms. Since one of the essential elements in memory is the fixation of a representation and this in turn is dependent upon the clarity and distinctness of a perception, it is evident that disorders of consciousness must affect memory.

In those disordered states in which clouding of consciousness is the most prominent symptom, the memory for the events which transpired during the attack is more or less unclear.

Certain conditions known as "absences" or "obscurations" illustrate this clearly. A person may do considerable traveling, may carry out very complex acts during one of these attacks and later have no clear recollection of what transpired or only a very vague one. Sometimes this condition assumes very important medico-legal aspects, when, for instance, a crime is committed during the attack, or some contract is entered into. One sees these conditions most commonly in connection with epilepsy.

Extreme self-consciousness, or an unusually concentrated attention of the entire organism to some part of the body or to some real or imagined ailment, may be spoken of as a form of hyperconsciousness. Dissociation of consciousness is an underlying disorder in many mental diseases.

2. Disorders of Perception

When a sensation is elaborated by the higher nerve centers and becomes a phenomenon of consciousness, it becomes a perception. Perceptions may be disordered in the following ways. First, the perception may be inadequate, not definite, or clear enough, as is seen in states of confusion or marked depression, or absorbing anxiety, etc. The patients do not perceive external impressions clearly. Things in the environment are vague, uncertain, confused. There may be, of course, varying degrees of insufficiency of perception, and as we have already seen, the individual may not even respond to the strongest stimulations when in a profound stupor.

The next type of disorder of perception is a mistaken or inaccurate perception or what is known as an "illusion." All the senses are subject to illusions, and normal people occasionally experience illusions of perception. In the normal, however, the aid of reason and of the other senses makes possible a correction of the deception and thus conduct is not controlled by it.

In the insane, on the other hand, the illusion may have the value of reality and controls conduct. The patient suffering from delirium tremens is terrified by the horrible animals which some indifferent object may cause him to perceive. One meets with illusions in various forms of mental disorders, but particularly in those due to toxic conditions and in the deliria of infectious diseases. Illusions as well as inadequate perceptions are due to a disorder of the ideational and associative processes. Wrong associations are formed which distort the image.

Hallucinations or imaginary perceptions is the third type of disorder of perception. The simplest definition of an hallucination is that it is a perception without an object. It is wholly imaginary. As is the case with illusions, all senses may be affected. Hallucinations are very common in mental disorders and exercise a marked influence upon the patient's conduct. The degree of this influence depends upon the stage of the disease. As the condition become chronic, and especially as enfeeblement of the intellect sets in, this influence subsides. The more intelligent the patient the more concerned will he be about the origin and source of an hallucination, and rather than admit that it is an evidence of mental trouble will endeavor to muster all sorts of bizarre explanations in defense of the reality of the perception.

Patients also take from the current ideas, notions or fads of the times, explanations for their hallucinations. One no longer hears patients attribute their experiences to diabolical influences, but in place of these he resorts to *x*-rays, wireless, etc., as explanations for the origin of the voices.

These hallucinatory experiences force themselves upon the patient and fix his attention. One sees patients walking about with their ears stuffed full of cotton in an endeavor to exclude the unpleasant voices. Or they stuff up every crack and crevice in their rooms to keep out the imaginary poisonous fumes and odors.

Hallucinations may affect very materially the emotional state of the patient in accordance with their content. The hallucinations may be pleasant, or extremely disagreeable, but in the final stages of certain disorders, the patients tend to become indifferent to the hallucinations. The disagreeable hallucinations are the most common, and sometimes so intensely affect the emotional state of the patient as to lead him to suicide or homicide, the act being carried out sometimes in response to a command of the imaginary voice. The extent to which hallucinations control the will depends upon the degree of insight the patient has into the pathologic nature of these false perceptions and upon the extent to which they affect his emotions.

The response to the hallucinations may be a very impulsive one, especially when hallucinations are accompanied by clouding of consciousness, or it may be a deliberate and carefully planned act. The patient who hears threatening voices, or the plotting of his enemies to do him harm, may deliberately set about to elaborate a counter-plot which he carries out with care and precision. One or more of the senses may be affected at the same time.

Hallucinations may be detected either through observation or through questioning the patient directly or indirectly.

The patient may be observed in a listening attitude, or may be heard conducting a conversation with imaginary voices, or he may be observed carrying out an impulsive act and when questioned will state that he did this in response to a voice.

The patient may complain about the annoying voices or odors or sensations, or he may be very reticent and suspicious about them. Sometimes the diagnosis must be made in spite of the patient's denial, and this is extremely difficult.

We shall not enter here into a discussion of the origin or purpose of the hallucination, beyond stating that where thorough analyses of hallucinations have been made, their purposive nature has been definitely determined and in some cases at least have been shown to be the expressions of a wish. Yes, even such terrifying voices as those which threaten to cut the patient to pieces may be projections into the environment of wishes which come back to the patient in the form of voices.

Of all the hallucinations the auditory are the most frequent and most important clinically. These may be simply noises, such as whistling, groaning, ringing of bells, etc., or they may be distinct words. The effect of the former depends upon the interpretation which the patient gives to them. The latter may consist of single words or of complicated discourses. One of my patients, a very able lawyer, would argue for hours with voices upon most technical and obstruse subjects. He used to get extremely excited and profane during the arguments and although clear mentally and able to carry on an intelligent conversation, he could not be persuaded of the pathologic nature of these voices. When I first knew him he had been in the hospital for 12 years, markedly hallucinated. Another of my patients, an extremely well-educated and intelligent physician, who was also an accomplished artist, insisted that he was being carried out of his room every night by pigmies who took him down town and exposed him to all sorts of indecencies. In the morning he would point to various imaginary spots on his bed linen as evidences of the experiences of the night before. This same man had spent some 40 years in an English prison for insane convicts after having killed a man in response to an hallucination.

Although hallucinations of hearing are present in many mental disorders, whenever present, especially when well developed and complicated, they indicate a bad prognosis. Of less importance are hallucinations of sight. They are usually found in toxic conditions and in deliria.

Other hallucinations met with are those of the sense of taste and of touch, of the temperature sense and of the sense of pain. Of particular interest is the "crawling sensation" seen in cocain habitués.

3. DISORDERS OF MEMORY

The next group of symptoms met with in psychiatry have to do with disorders of memory. The process of memory involves three operations, namely, the fixation of a representation, its preservation and its recall in the field of consciousness with the recognition as having taken place at some previous time.

Disorders of memory may affect these processes singly or together and we may therefore have different varieties of amnesias.

The amnesia may be due to a faulty fixation owing to an indistinctness of the perception. Hence all conditions which are accompanied by a clouding of consciousness will cause this form of amnesia. The processes of conservation and recall may be intact but there is nothing to conserve or recall. But an additional element enters here besides the distinctness of perception, which has to do with the fixation of the impression.

Amnesia may be due to a faulty functioning of the process of conversation. Of course, this is a relative term. A man may have a very poor memory and still be entirely normal. But the loss of this capacity to retain is one of the most characteristic manifestations of the organic group of disorders.

It is not infrequently the first symptom of such deteriorating processes as senile dementia or paresis. There is also possible a purely functional memory defect, again a purposive mechanism and you will find it not at all difficult to appreciate the dictum that we forget things which we do not wish to remember. But we are speaking here of organic defects of memory. Finally an amnesia may be characterized principally by a defect in recall. This type of defect is encountered most frequently in the acute psychoses and has a good prognosis.

Amnesia may be partial or complete, it may be circumscribed and limited as to the period in the patient's life involved or as to certain experiences. Thus one of our patients had a complete amnesia for a certain series of events during which arose the conflict which led to the development of an hysteria.

Of considerable interest are the various falsifications of memory. This may either be in the form of an illusion or of a true hallucination. A past event may be recalled in an altered form, which may entirely change the import of the event. It is characteristic of a certain type of lying in psychopathic individuals or even in normal people. We are all familiar with the modifications which a story assumes when retold by a number of people. Experiments have demonstrated that sometimes the original story has entirely lost its true character. This is not, of course, due to conscious lying. But it is very significant as illustrating the relative value of hearsay evidence. Children and the weakminded are especially subject to illusionary reproductions. It is also seen sometimes in the senile dement. An hallucination of memory is the recall of something which has never occurred.

This is at the basis of that strange phenomenon known as "pathologic lying," a not infrequent manifestation of constitutional inferiority. A report of two very interesting cases of this nature is given in my monograph on Forensic Psychiatry.

Pseudo-reminiscences are not uncommon in the insane and feebleminded.

The further consideration of disturbances in thinking embraces the disorders of attention, ideation and judgment.

4. Attention, Ideation and Judgment

Attention may be spontaneous or voluntary and deliberate. The former is merely the spontaneous, undirected attention to circumstances, while the latter is more purposive and deliberate and directs the flow of thoughts, it is part of the voluntary conscious dsychic activity. The conditions of disordered consciousness which we have already considered are accompanied by a reduction of the faculty of attention to the degree in which consciousness is reduced. Spontaneous attention is part of consciousness. We are more concerned with disorders of deliberate attention, disturbances in the stream of thought.

The most commonly met with is an instability of attention, an exaggerated mobility. The individual's attention can be gotten but cannot be held. We speak of this condition also as an abnormal distractibility. Nothing which comes within the individual's horizon distracts his attention. It is evident that this type of attention disorder must interfere materially with the voluntary association of ideas, that is, association which is controlled by a guiding idea, since one of the principal functions of deliberate attention is to prevent automatic associations from obtruding themselves upon associations formed under the control of a guiding idea. Voluntary associations become sluggish, delayed and when combined with disorders of consciousness constitute the picture of confusion.

Disorders of attention and association give rise to two phenomena frequently met with in mental disorders, namely, flight of ideas and incoherence. The one is characteristic of true manic states, the other of dementia precox. In incoherence, ideas follow each other without any connection, while in flight of ideas there is some relation but a very superficial one. Here, too, analytic psychology has shown that what appears to be a meaningless productivity, really has some meaning if we only take the trouble to search for it. Sometimes the two symptoms appear in the same patient, depending upon the degree of disintegration. Other disorders of ideation are fixed ideas and imperative ideas. The difference between the two is that a fixed idea harmonizes with the remainder of the mental setting, it is never recognized by the patient as something foreign to him, while an imperative idea is appreciated by the patient as something foreign to him, he fights against it, wishes to be relieved from it and has insight into its pathologic nature.

A very common manifestation of mental disorder is the delusion. A delusion is a false judgment maintained in the face of obvious evidence to the contrary. A pathologic delusion is one out of which the holder cannot be reasoned out. A delusional state of mind is where judgment is so impaired as to habitually lead to wrong conclusions in spite of contradictory evidence. Sometimes this leads to the building up of a well-organized delusional system which in extreme cases leads to the entire transformation of the personality. Such a well-organized system of delusions may possess a good deal of plausibility, and it is for this reason that very dangerous paranoiacs sometimes escape detection for years. On the other hand, the various delusional ideas may have no connection, one with another, and we speak of them as incoherent.

The degree of coherency of the delusional system, the degree to which it is in harmony with the emotional tone of the patient, and the extent to which it affects his actions are very good criteria upon which to judge the degree of mental dissolution present.

An advanced case of dementia may, for instance, express very bizarre delusions of grandeur, and at the same time will gladly carry out menial tasks which are wholly out of keeping with the person he imagines himself to be. He will declare that he is an emperor while perhaps sweeping the floor. He may recite most distressing delusions of persecution and do so indifferently or perhaps exhibit the opposite emotional tone to that which the delusion in question would be expected to provoke. His delusions are also likely to be disconnected, incoherent and changeable from day to day.

But the degree of coherency of a delusional system does not always bear a strict relation to the seriousness of the malady, or to its probable outcome. In true paranoia, perhaps the most serious and certainly the most dangerous form of mental disorder, the delusional system is regularly well systematized and coherent and the patient shows very little, if any, disintegration.

As a rule, and particularly during the early stages of a mental disorder, the delusions are in accord with the emotional tone of the patient and have a great capacity for controlling his conduct. A patient suffering from depressive delusions will be melancholy and may attempt suicide.

Delusional ideas may be divided, in the main, into depressive, expansive and persecutory.

Depressive ideas are the outstanding symptoms of certain forms of mental disease and are part of the clinical picture of a good many, especially during the early stages. The patient may show a tendency to self-depreciation and undue humility, will declare that he does not deserve the food he eats or the clothes he wears, or the attention of his friends; that he is worthless, a wretch, a criminal, etc. Sometimes the self-accusatory ideas become more specific and the patient will declare himself responsible for the death of a relative or for some other general calamity, such as fires, floods, earthquakes, etc. Sometimes they consider themselves guilty of "the unpardonable sin," they are doomed to ruin, there is no hope for them, in fact they are to be tortured and executed that night, etc. They will complain of various imaginary ailments, at times even declaring they have no stomach or liver, or that they have actually wasted away, are nothing, etc. The patient suffering from such depressive ideas is melancholy, anxious, fearful and genuinely suffers very much psychical pain. He may refuse to take nourishment or may commit suicide. In his bearing he is very much dejected and humble, craves assurance and sympathy, but the delusional ideas control him entirely. Especially distressing is the clinical picture known as "involution melancholia" which develops in some people in connection with advancing age.

The opposite picture to this is seen in patients exhibiting delusions of grandeur or expansive ideas. Ordinarily these ideas are more bizarre in their exaggerations and accompany states of more or less profound mental disintegration. Thus the ideas are ordinarily out of keeping with the actual bearing and attitude of the patient. The man who imagines himself to be an emperor or God will submit indifferently to the restrictions of the hospital, etc. The ideas which they express are sometimes bizarre and wholly incompatible with reality.

A patient of mine, a paretic, came to Washington to sell the United States Government forty million carloads of axle grease. The same patient declared that he lived in a diamond palace and that he was 40 yd. in height. When less demented, these patients endeavor to act the part more or less in dress and demeanor. They will decorate themselves with all sorts of phantastic decorations, will keep themselves aloof from the others and constantly issue commands to those about them or threaten them with an instant annihilation.

In acute manic states with much excitement and activity, the grandiose delusions may give direction to the patient's actions. Of much interest is the association of delusions of grandeur with persecutory delusions as seen in paranoia. Here, it would seem that there comes a stage in the patient's disorder when he begins to seek an explanation for being the object of so much persecution and attention, and finds the explanation in the belief that he must be a truly unusual person, someone very important, indeed, to attract so much attention.

Ordinarily persecutory ideas are of a very painful character, but unlike the melancholic, the persecuted person aggressively protests and fights against the persecution. The development of these ideas is, as a rule, gradual, the patient becomes suspicious, sees further and hears further than anyone else, everything in the environment is given a special meaning and is misinterpreted as having reference to himself and as being directed against him. Gradually more definite persecutory ideas are developed and more and more people become involved in the plot to destroy him.

Sometimes the persecutory ideas are the result of the content of hallucinations. The patient hears voices accusing him of various crimes or horrible misdeeds, not infrequently of sexual perversions. The voices begin to threaten him, etc. When accompanied by hallucinations, the persecutory delusions are more apt to control the patient's conduct and to lead to impulsive action, sometimes homicide or suicide. Ordinarily the patient has but a vague idea at first of the source of his persecutions. He will speak of his persecutors as "they." When the delusions become more systematic the patient has usually in mind some person or persons whom he considers responsible for his persecutions.

5. Disorders of the Emotions

The next disorders which we will consider are the disorders of the emotions. Some degree of emotional disturbance constitutes part of every form of mental disorder. But in some of these conditions the emotional disorder is the outstanding feature of the difficulty.

The first of these is emotional indifference. This is a prominent symptom in certain disorders, particularly in the terminal stages. In dementia precox, on the other hand, it may be prominent at all stages of the disorder, and even in what is known as the predementia precox stage. We have already stressed the point of the importance of emotional indifference and dullness. In the life histories of certain patients we discern a peculiar lack of emotional responsiveness to situations, a peculiar obtuseness and awkwardness in emotional contact with others. Patients may have insight into this, especially in the beginning of the disorders, and will complain that they have become indifferent to things. A complaint of this sort should always be taken seriously since it is one of the first indications of a dissolution of the personality.

We have already spoken to some extent of the emotions of joy and sorrow which may go with certain delusional ideas. The more commonly met with are depressive states. In connection with depressions certain bodily changes take place which can be clearly observed. There is a disturbance in circulation, in respiration and in the digestive mechanism. The intense pain that goes with the depression diverts the attention to the extent of interfering with these functions. Another phenomenon which goes with depressions of a certain type is a sort of paralysis of the will and function. It is this extreme psycho-motor retardation which accounts for the fact that a greater number of melancholics do not commit suicide. They lack the energy and will to carry out the act. At other times the depression is accompanied by marked restlessness and agitation. The patient walks about restlessly, wringing her hands in an agitated, disturbed manner. This type is particularly characteristic of involution melancholia. The depressive states are frequently accompanied by delusions though this is not always the case.

Intense joy or pathologic euphoria, on the other hand, is accompanied by an acceleration of the mental and physical powers. Ordinarily it is accompanied by motor excitement, talkativeness, exaltation.

Morbid euphorias are seen in a setting of emotional exaltation, psycho-motor hyperactivity, flight of ideas, while depressions occur in a setting of emotional depression, psycho-motor retardation, difficulty of thinking.

Morbid anger or rage is most typically represented by the wild excitement of epileptics and others who are suffering from obscuration of consciousness. Sullenness, irritability and impulsiveness are accompanying symptoms and usually precede the outburst of rage. Some degree of anger may be part of the picture of most mental disorders, with the exception, perhaps of involution melancholia.

6. Disorders of Volition and Acting

We have to consider finally the disorders of volition and acting. In all psychoses or mental disorders there is some indication of a disturbance in acting. In fact the community cares very little what delusional ideas a man may entertain or what his state of feeling may be just so this is not expressed in some disorder of action. When the persecuted paranoiac reacts to his delusions, the objective evidence of an existing mental disorder comes to light. The disorders of will and action may express themselves in a number of ways. In the first place, action may become less and less deliberate and more and more impulsive.

The impulsive action may be in response to some emotional state or it may have no discernible reason for it. An excited manic may be pushed inadvertently by another patient and without stopping to consider whether or no an offense was meant, will impulsively strike the patient. Or some remark by someone within hearing may be taken as a personal slur and cause an impulsive reaction. On the other hand, in the midst of a calm conversation, the patient may suddenly tear up an article of his clothing or assault someone or begin to strike himself. This tendency to impulsive action frequently goes hand in hand with what is otherwise taken as a complete paralysis of the will. Certain patients, especially those suffering from what is known as catatonic stupor, may be for days or weeks absolutely immobile, mute, not even feed themselves or attend to the calls of nature, and then will all of a sudden, out of a clear sky, as it were, carry out an impulsive act, such as running up to the window and smashing the panes. Along with this paralysis of the will, almost complete abeyance of function, there goes sometimes a peculiar suggestibility, or on the other hand, a resistance and negativism.

The patient's limbs may be placed in any position, even a very awkward one, and they will be retained in this position indefinitely. This condition is known as waxy flexibility of "flexibilitas cerea." Or the patient may be in what is known as a state of "command automatism." Any acts performed by the observer are imitated by the patient in an automatic way—or anything said within his hearing is repeated in a monotonous automatic way. The patient on the other hand may be extremely resistive to suggestion. He cannot be made to respond to commands, not even to those which have to do with self preservation, such as taking food, etc., a passive negativism. Or he may be actively negativistic and do just the opposite of what is wanted of him. When told to open his eyes he will shut them, to sit down, will stand up, etc.

Another curious phenomenon of disorder of will is known as stereotypy. The patient seems to persist in retaining the same attitude or in repeating the same phrases, or in carrying out peculiar mannerism of walking or talking for considerable periods of time.

Now it is very likely that all these curious manifestations have some definite meaning to the patient, they serve some definite purpose, no matter how strange and unaccountable they may appear to us. Familiarity with the common manifestations of mental disorder renders possible to discern a tendency for the symptoms to group themselves in certain definite constellations. It is very helpful to keep in mind this grouping, since it enables one to discern more clearly meanings back of the psychiatric reactions of the patient.

Thus one might speak of the psycho-neurotic reaction type in which the underlying process seems to be a breaking up of the unity of the personality, a splitting of the personality. Some constellations of ideas which are highly charged emotionally make a strong and persistent claim for attention, thus diverting to themselves a greater or less portion of the personality. In extreme cases we have a complete splitting and we are dealing then with a double or multiple personality. Then again it is possible to speak of the dementia precox type of reaction in which the underlying mechanism is a tendency to withdraw from reality and the objective world into a world of one's own creation, a phantasy world in which delusions and hallucinations take the place of actuality and in which cravings which are incompatible with reality are realized. Then there is the depressive type of reaction in which the individual appears to have succumbed to the strugggle, gives in, becomes retarded in action, in body and mind and tortured by the thoughts and feelings which were at the bottom of his conflict. In the maniacal excitement we see, on the other hand, an attempt at running away from oneself, a flight into reality with such pathologic acceleration of action and thought as to divert attention from oneself. It is an over-busying with the outside world which leaves no time or energy for concern with subjective matters. Then there is the paranoid type of reaction, in which delusions of persecution and grandeur play the dominating rôle, and which in reality is a projection of a struggle with oneself into the environment, which then is apprehended as coming back from the environment in the form of persecutions, etc. The tendency in psychiatry is increasingly in the direction of seeking in the manifestations of the mental disorder some meaning as regards the life career and adaptive dispositions of the patient.

Earlier in this chapter we have indicated that the psychoses of childhood most commonly met with are, the toxic-exhaustion deliria and post-febrile psychotic states, the syphilitic disorders, principally, juvenile paralysis of the insane, the schizophrenic states and the manic-depressive disorders. We will briefly describe these conditions.

THE TOXIC-EXHAUSTION PSYCHOSES

Viewed from the point of view of causation, these disorders definitely belong in the exogenous group, in that here clearly an external agent in the form of some pathogenic organism brings about the altered mental state. It is not altogether clear how this takes place, especially is this true of those disorders which do not lend to clear-out organic changes in the nervous system.

Nevertheless, even in these purely exogenous disorders the element of personality plays an important rôle in determining the clinical manifestations. Perhaps also more weight should be given to the question of predisposition to pathologic reactions to infection. Clinically it is pretty well established that the severity of an infection is not the sole determinant whether or no a given individual will react to it by some psychopathologic disturbance.

The situation is well illustrated in the acute pathologic reactions which some constitutionally inferior individuals manifest in connection with the ingestion of relatively slight quantities of alcohol. The extreme intolerance for this noxious agent is reflected also in a more generalized intolerance to assaults of any kind upon the organism.

In dealing with adults some of the outstanding disorders met with in this group owe their origin to various habit forming drugs, to metal poisoning in connection with deleterious occupations and various gas poisonings. Naturally this does not apply to children though it should be noted in passing that the use of heroin is becoming increasingly common among young adolescents in the larger cities. For our purposes the conditions to be considered here are those which develop in connection with some acute somatic disorder, the exanthemata and other acute infectious disorders of childhood.

In connection with an acute disorder of this sort the child may develop a delirious or acute confusional state with an extremely varying clinical picture. The delirium may be marked by severe motor excitement and incoherence of utterance, or by multiform hallucinations with deep confusion or a dazed, bewildered condition. There may also be manifested epileptiform seizures, catatonic-like symptoms, stupor and impulsive reactions. These delirious states may occur during the prodromal stage of the somatic disorder, during the height of the fever or during the period of defervescence when it is in the nature of a collapse delirium.

In connection with convalescence from acute infectious disorders especially when the seige has been a particularly trying one, patients may develop various reactions of a psychopathologic nature, such as depressions, irritability, a tendency to misinterpret fairly ordinary events and some confusion. These post-febrile disorders undoubtedly have as etiologic factors the physical asthenia and exhaustion of the patient, whereas in those occurring during the course of the somatic disorder, some transitory nutritional or metabolic disturbance might be held accountable for the abnormal mental manifestations.

Apart from these disorders which occur in connection with or immediately in the wake of an acute somatic disease, mental disorders due to pure exhaustion, either from severe hemorrhage, unusual physical exertion, protracted loss of sleep and inanition are sometimes seen in children and young adolescents. The clinical picture in all these conditions is that of a delirious or acute confusional state, although towards the latter part of the disorder, more definite paranoid manifestations may be present. During the acute state a greater or less degree of peripheral analgesia is often observed and occasionally a narrowing of the field of vision.

The condition most commonly lasts three or four months, recovery is usually complete although as regards adults it should be pointed out that a certain percentage, perhaps as high as 10 per cent. suffer from a permanent mental reduction of greater or less severity. It is very difficult to be specific about the prognosis in any given case during the acute stage of the disorder, when mental confusion is very marked. Indeed, it is sometimes very difficult to differentiate some of these conditions, in which catatonic-like symptoms are present, from catatonic dementia precox, a source of error which very determinedly affects the nature of prognosis one should render. The treatment, although occasionally feasible in the patient's own home, has to be carried out usually under the auspices of a hospital for mental disorders. The patient's confusion and disorientation, the tendency to impulsive reactions and misidentification render treatment in the patient's home not only difficult but sometimes dangerous. During the acute stages, rest in bed, forced feeding and the avoidance, as far as possible, of overstimulation to the mind, are the outstanding features of the treatment. Occasionally the patient's extreme restlessness and motor excitement have to be managed by restraint. To this end the various forms of sedative hydro-therapy should be resorted to, though the possible exhausting effect, for instance, of the continuous bath must be kept in mind. Occasionally mild sedative drugs must be resorted to to assure for the patient the necessary amount of rest and sleep. During convalescence care should be taken against too abrupt physical or mental exertion, since this may lead to a relighting of the process.

For a very excellent treatment of the nature, causes and management of these disorders, the reader is referred to Stoddard's "Mind and Its Disorders."[15]

Of special importance in connection with this subject are the numerous and varying abnormal mental and behavior manifestations in children which develop as part of or as a sequel to lethargic encephalitis. It would lead us considerably beyond the scope of this paper to enter into a detailed discussion of this subject here. But the fact that lethargic encephalitis has very likely become endemic in this country, and that in such a large proportion of cases the condition is followed by mental aberrations, renders this subject extremely important for the pediatrician.

The numerous efforts that have been made to classify these conditions in detail seem to the writer considerably forced. At any rate nothing of practical value is gained from this and very likely clinical experience does not justify a complete and final classification at this time. In view of the manifold brain lesions discovered at autopsy it is difficult to see how an absolute classification is possible at all. The disease belongs essentially in the category we have just discussed with the important difference that so commonly the symptoms seem to be based upon meso- and ecdodermic changes in the brain. The mental picture of these children as given by Leahy and Sands[16] is characterized by a purposeless, impulsive motor hyperactivity, marked irritability, definite attention disorders, distractibility and changing, variable mood, inadequate and inconsistent emotional reactions, marked insomnia and feelings and intense eroticisms.

JUVENILE PARALYSIS OF THE INSANE (JUVENILE PARESIS)

Apart from the fact that this condition is encountered with increasing frequency in children, its inclusion here is justified also on the score of the rather considerable possibility of mistaking it for the more common and decidedly less grave defect states in children. I have seen two cases of juvenile paretics in institutions for the feebleminded in whom the behavior manifestations were wholly misunderstood because the existence of paresis was not recognized. Paresis is a syphilitic disorder developing most commonly 10 to 15 years after the initial infection. Juvenile paresis is a sequel of congenital syphilis, developing usually in the second decade of life, although cases have been reported at the age of seven.

When fully developed the clinical manifestations do not differ materially from those observed in the adult paretic, except that the mental symptoms are apt to be predominantly those of a profound defect state. Before these children have achieved the degree of mental development commensurate with their physical age, the destructive lesions of the nervous system not only check normal development but rob the individual of some of the equipment already attained.

Remissions, if present at all, are much less common in juvenile paresis, than in the adult form of the disorder and the progress to dissolution is apt to be more rapid than in the adult cases. Otherwise there are no significant differences in the two types. The symptoms, course and treatment of this condition are so well known as to require no restatement here.

THE SCHIZOPHRENIC REACTIONS (DEMENTIA PRÆCOX)

Perhaps in connection with no other mental disorder is it as significant to make an early diagnosis as in connection with dementia præcox. This is due to the growing belief on the part of psychiatrists that this disorder is perhaps best understood on the basis of a progressive habit deterioration. If this is correct it is of utmost importance to determine, as early in the life of the individual as is possible, what the conditions of life are and what the characteristics of personality which give rise to these unhealthy reactions and to the habituation to them.

There are still a great many points at issue as regards causation as well as to what constitutes the essential clinical features of this disease. We cannot enter here into a detailed discussion of these points, except to state that more and more evidence is being accumulated to the effect that often personality traits can be distinguished in the child which presage the development of a dementia præcox process.

These characteristics refer more particularly to the instinctive-emotional development of the child than to his intellectual development. Indeed, the future dementia præcox patient may be exceptionally bright as a youngster, but is apt to have considerable difficulty in adjusting himself to the demands of school life. As a child he is apt to keep by himself, does not seem able to get on common ground with other children; spends much time in absorption in his own day dreams and fancies and resents being called out of them; is apt to be sensitive and to take knocks and disappointments as slurs or injustices. These children find it particularly difficult to frankly externalize their thoughts, to be natural in their intercourse with other children.

The frank mental disorder rarely manifests itself before puberty, but with the onset of this period clear indications of a beginning mental aberration may come to light. At first there may be present what seems to be merely an accentuation of an already existing tendency to shyness and withdrawal, but in the great majority of instances the early manifestations of this disorder are already characterized by a rapidly progressing emotional apathy and general queerness of attitude.

The moods these young patients exhibit are apt to be out of accord with the real situations surrounding them, and from the very first they may exhibit bizarre mannerisms of speech and action.

The later development of the disorder may tend especially in the direction of any one of several types that have been differentiated, but most frequently one deals in the young with the hebephrenic form. Commonly the development of this form of dementia præcox is insidious in nature. The child may have been previously quite normal in all respects, a good scholar and exhibiting a lively interest in things. Gradually his interest in school work begins to wane, he loses ambition in his work, becomes negligent about his personal appearance and the various social amenities to which he may have paid a good deal of attention before. When spoken to about these changes by parents or teachers, he may respond with a meaningless, indifferent sort of shrug of the shoulder, or laughter, or by an impatient irritability and moroseness. The characteristic which will force itself upon the attention of those who observe him is his profound lack of concern over it all.

The early hebephrenic may exhibit intense enthusiasm but these are unfortunately very momentary, he soon loses interest and will, and sinks back into a distressing apathy. Along with this there may come to light an absorption in fancies and day dreams which serve more and more to satisfy these youths in place of active attention to the immediate demands of daily life. Not uncommonly there is a flare up of religious pre-occupation, or an interest in fads of one sort or another with regards to food, health, personal habits, etc. Memory is retained and orientation is unimpaired for a long time and nothing further may come to light of a pathologic nature. The loss of interest, ambition and will leads many a former high school and even college student to drift gradually into some simple, menial occupation which carries with it no responsibility and which he can change frequently on the impulse of the moment without calling forth much criticism.

The situation is particularly distressing when this occurs in a well-to-do and refined family with high ideals and ambitions for their children. The youthful hebephrenic may not exhibit, at any rate, not for a long time those frank symptoms of mental disease as delusions or hallucinations which might render the seriousness of his disorder obvious to everyone and gain for him the benefit and protection of a hospital for the insane. Thus, for a long time it may be very difficult and even impossible to convince his relatives of the true nature of the difficulty and many a parent, even among intelligent people, insists upon dealing with the problem in a purely disciplinary manner when his child should receive treatment in a hospital.

These patients lack wholly in insight into their condition, another element which aggravates the situation. When the protection of proper connections, family or money is lacking, these youthful hebephrenics soon drift into a life of vagabondage and total abandonment, and are apt to become drug addicts and alcoholics, and to find their way into a life of petty criminalism.

I cannot stress the point too strongly that in the history of practically every early hebephrenic one is able to discover an intense preoccupation for a greater or less period of time with problems of sex.

Usually excessive masturbation with considerable play of phantasy in connection with this indulgence, in which, often, near relatives of the opposite sex play the rôle of a sex object, comes to light. In some cases an intense moral conflict, with profound self-accusatory and self-depreciatory ideas accompanies the situation. Often fancies of a homosexual nature occupy a prominent place in the picture. This is the simplest form in which the disease may manifest itself.

Even in less pronounced manner than the above description, the disease may creep upon an early adolescent. It is important to keep this clearly in mind in order to avoid misunderstanding and mishandling the case. In a certain percentage of cases no further deterioration takes place and the patient may find in some simple and non-taxing occupation, a fairly comfortable solution to the problem. In rare instances there may even take place some degree of re-awakening of the patient's former interest and enthusiasm.

The disease may set in with a good many neurasthenic manifestations, such as headache, insomnia, irritability and excessive fatigability, and it is important not to miss the more profound disorder which these symptoms cover. The onset and course of hebephrenia may, on the other hand, assume much more stormy characteristics, so that even the lay person recognizes readily the seriousness of the situation. From the very beginning the changes in character may be accompanied by intense emotional outbursts, delusions and hallucinations. Among the delusions self-accusatory notions of a rather bizarre character are apt to be prominent. Occasionally the anxiety accompanying these thoughts may lead to intense reactions, even suicidal or homocidal attempts.

We cannot enter here into a more detailed description of the hebephrenic disorder. The main points we wish to stress relate to the milder forms of this disease where indications of a very serious and usually chronic mental disorder may be mistaken for the simple moodiness of the adolescent boy or girl. Not only do educators and even physicians fail to keep this possibility in mind, but they discourage parents from viewing the problem with the necessary degree of seriousness.

The hebephrenic form of dementia præcox is by far the most frequent form in young adolescents. Occasionally a young boy or girl gives the first indication of developing dementia præcox through an acute catatonic episode. We have seen these episodes in children as young as 11.

The prognosis is always grave in dementia præcox and the treatment difficult and protracted. It is to be strongly emphasized that treatment should not be undertaken except by those thoroughly experienced in dealing with these patients. This caution is especially indicated since the psychoanalytic approach to these problems is being more and more encouraged in some quarters. I have often seen cases of young hebephrenics whose condition has been distinctly aggravated through an unwise deep probing into their mental content.

One of the outstanding aims of treatment is to assist the patient to a better insight into his condition. It is particularly desirable to point out the unhealthy character of his method of adaptation to the problems of life. Attention must be devoted to the discovery of the occupation which has the capacity to call out and hold the patient's interest. The treatment, of course, differs in many essentials in the individual case, since the patient may be brought to the physician's attention in a condition which renders treatment in a hospital imperative. But experience leads one to the opinion that once this diagnosis is established the patient has the best chance when placed, at least for a brief period of time, in a hospital where he can be closely observed and studied and where future plans of treatment can be determined with greater accuracy.

Only in exceptional cases should even a trained psychiatrist undertake the treatment of one of these patients without the kind of careful preliminary study which only a modern hospital for the insane can carry out.

THE MANIC-DEPRESSIVE PSYCHOSES

The very familiar mental disorders of adults which fall under this category are occasionally observed in children, principally in young adolescents. The manic-depressive disorders are particularly characterized by periodicity and alternation, the patients usually fully recovering from the single attack but strongly liable to recurrences. The single attacks may be either those of mania, in which the outstanding symptoms are emotional exaltation, motor excitement and flight of ideas; or of melancholia, the opposite picture, in which emotional depression, retardation of thought and action and difficulty in thinking are the outstanding features; or the central picture may be that of stupor. Delusional ideas, usually in accord with the dominant emotional trend are present, and mixed states in which various combinations of the central symptoms mentioned above also occur.

Commonly one is dealing in these patients with an unstable mental equilibrium, the first attack usually occurs in the third decade of life. Recovery from single attacks after four to six months is practically the rule, followed with remissions of greater or less duration and a strong tendency to recurrence.

The reason we include this disorder here is that it has to be differentiated, albeit in rare instances, from the profound emotional fluctuations of the psychopathic child. In the latter, as has been indicated elsewhere, deep depressions and pronounced excitements may develop under stress which ordinarily disappear when the stress is removed. In the manic-depressive disorder the occurrence of an attack is largely and wholly determined by inner conditions. The prognosis of the single attack, as has already been indicated, is favorable. Treatment has to be carried out under a hospital regime in practically all cases.

FUNCTIONAL NERVOUS DISORDERS OF CHILDHOOD

Perhaps no phase of childhood psychopathology offers as many difficulties of understanding and specific differentiation as that which is embraced under the rather broad designation of "nervous child." What does one mean by nervousness in the first place? What is this condition based on? How much of it is modifiable by treatment?

These questions, often enough quite unclear when dealing with adults, are even more obscure in connection with childhood difficulties. In the developmental histories of children brought to us on the complaint of nervousness, certain facts repeat themselves with such regularity that it might be well to begin our presentation by a citation of these facts.

In the first place mothers tell us that from the very moment of birth these youngsters seemed to be extraordinarily sensitive to ordinary sensory stimuli. These children cannot endure bright light, exhibiting discomfort, irritability, and often outbursts of crying in the presence of moderately strong light. They are very sensitive to the quality of clothing with which their bodies come in contact, being fidgety, restless and sleepless when their clothing happens to be a little rough. Noises, even of moderate intensity, such as the creaking of a door, or sudden dropping of a hard object, or even the voice of an adult, if at all gruff and loud, startle them violently and may even bring on a fit of crying. They are unusually sensitive to changes in temperature and the daily toilet, unless very carefully adjusted, throws them into a panic. All this extreme sensitiveness to sensory stimuli is reflected in their sleep habits. They often wake with a start, crying loudly and much, and frequently give external evidence of fear and anxiety. Occasionally involuntary twitchings of various groups of muscles are observed as well as an intensity of emotional display in connection with a crying spell. Nutrition is usually, though not regularly, bad, and the reaction to the ordinary ailments of childhood is very intense.

It is this exaggerated sensitiveness which manifests itself from the earliest infancy which lends much strength to the contention that the nervous child carries a constitutional burden primarily. As these children grow older this hypersensitiveness, and one might say, over-awareness, is seen to be coupled with an exaggerated fatigability. Even before formal demands upon the child's energy are made, as in connection with school life, this heightened fatigability is discernible. Commonly one encounters much difficulty in getting these children to sleep, many of them spending much time in a restless endeavor to combat an undue awareness and wakefulness, until exhausted, they finally drop off to sleep. But they cannot wake in the morning, feel tired and exhausted, spend much of the day in a peevish, irritable state and by the time they are ready for bed they are so worked up that sleep again eludes them.

Much of this is due solely to fatigue and can be relieved to a very large extent by appropriate measures for the elimination of over-stimulation and the provision of rest periods during the day.

As the child grows older and contacts with its environment become broader, new factors enter the situation which complicate matters materially. In the first place there may be added to the child's original handicaps of hypersensitiveness and heightened fatigability, the decidedly bad effects of unwise training and management. Apart from the common fallacy of seeing in the nervous child's over-awareness and over-sensitiveness only the signs of brightness which commonly lead to over-stimulation by deliberate forcing of his development, unwise management may lead the child to a kind of exploitation of his innate difficulties which is responsible for many of the behavior problems of the nervous child.

Thus headaches, vomiting, insomnia, which originally may be purely the expression of the child's inferior nervous organization, assume through unwise management new qualities for the child and are exploited to his own detriment and to the despair of his parents and teachers. This preferential position in the household and classroom which the nervous child may come to seek is not the worst of the by-products of his original nervousness. It also has the capacity of instilling a very deleterious sense of inadequacy and inferiority which materially hampers later adaptation to life.

The conditions which determine these unhealthy elaborations of original nervous tendencies have been discussed in detail in the introduction to this chapter.

Other factors which complicate the situation of the nervous child are the manifold expressions of a more psychic character which develop by preference upon the soil of original nervousness.

Thus, no matter how far one might go in attributing the various psycho-neurotic manifestations of childhood to life experiences of a deleterious nature, the nervously dispositioned child is more apt than the normally endowed child to develop these reactions. Thus one sees not uncommonly in nervous children clear-cut instances of anxiety states, compulsive thoughts and actions, phobias of a specific kind and a more generalized phobism, quite well-marked instances of conversion hysteria, particularly common being hysterical vomiting, tics and spasms. Then the related varieties of functional speech disturbances which should always be considered as being primarily psychoneurotic manifestations.

Another series of phenomena owe their presence solely to a difficulty which the nervous child experiences in outgrowing some of the characteristics of the infantile state, such as thumb-sucking, bed-wetting or ready resort to crying and tantrums when balked in his desires.

It is important to note, however, that all of these manifestations, although developing by preference upon the soil of a constitutionally poorly endowed nervous system, have in addition specific causes for their existence. In the treatment of these disorders, it does happen occasionally that mere attention to the general problem of the underlying nervousness, such as simplifying the child's environment, removing of obvious environmental irritations, and attention to the child's general health clear up the situation. But commonly there is needed, in addition, a definite management of the specific difficulty.

The common causes of the difficulties have been stated in the introduction to this chapter. It is not necessary in all cases to assume an underlying constitutional defect. The child who is over-stimulated by over-ambitious parents; the child who is exposed to continuous failure at school; the child who because of a troublesome mental content becomes anxious and self-accusatory, all of these may develop the common manifestations of a nervous child without any underlying constitutional defect.

In the management of the nervous child, no matter what the manifestations may be, there are certain general principles which must be followed. Firstly, a careful hereditary and developmental history must be gained to determine if possible to what extent the innate factor is operative in the case. Of special significance is a careful account of the time at which the various outstanding points in the child's development took place, such as teething, walking, etc., and of the manner in which the child reacted to the events in his development. One of the best criteria for the determination of the possible degree of innateness of the nervousness is the child's sleep history and the history of his reaction to the common diseases of childhood.

Secondly, a careful physical study of the child with the view of eliminating a possible physical irritant at the basis of the nervousness. We have seen children improve markedly and even entirely become rid of their nervousness upon the correction of some special sensory defect, or a malnutrition, or an anemia or some parasitic disorder.

Thirdly, a careful study and analysis of the child's mental content. Ordinarily a very simple procedure in young children, this becomes quite difficult with older boys and girls unless all conditions are entirely favorable. Among these conditions, the attitude and manner of the examiner are of prime importance. It is remarkable how a single opportunity for freely talking out a troublesome mental content may relieve a child of a troublesome nervousness.

Fourthly, a thorough acquaintance with the adults surrounding the child. When one goes beyond the purely physical and physiologic factors at work in the nervous child and considers the child's entire personality, the importance of this becomes obvious. It is for this reason that the mere removal of a child to a new environment sometimes produces remarkably beneficial results.

More specific directions for the management of the nervous child cannot be given here. It must be clear that although the concept of the nervous child is widely employed as though it were a specific entity, practically every case presents a different problem and requires an approach appropriate to it. It might be added, in conclusion, that there is ample evidence to the effect that just as good results and better are obtained from an approach which is mainly psychologic as are from the more traditional therapeutic endeavors by means of drugs. The reader is referred for a more detailed elucidation of this subject to the excellent book on the "Nervous Child" by H. C. Cameron.[17]

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CHAPTER CLXXXVI

DEFECTS OF SPEECH OF CONGENITAL, OR DEVELOPMENTAL ORIGIN

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THE PHYSIOLOGIC FOUNDATIONS OF SPEECH

The child is the mature man, or woman, in the process of making. The physiologic developmental processes of the infant, if continued normally through a sufficient number of years, lead to a normally constructed adult, both as to mind and body. Granted that the physiologic basis at birth is normal for both mind and body, and granted that at every step the natural development continues normally, the normal man is the inevitable result. It is from this standpoint of the normality, or abnormality, of the mental and peripheral bases of speech in the infant and the normality or abnormality progress of development of function, that one must view the subject of disturbances of speech in childhood. For nearly all disturbances of speech in childhood are dependent either on fundamental abnormality, or on abnormality occurring in the processes of development themselves. In brief, nearly all disturbances of speech in childhood are congenital, or developmental, in origin.

Here, then, lies the keynote to the natural manner of approach to the subject of defects of speech. Unless one understands the foundations of speech, he is quite unable to understand defects of speech. If one does understand the normal foundations of speech and does understand the manner in which thése foundations may become disturbed in the process of development, he then becomes capable of understanding any perversion of speech.

The extraordinary complexity of this function, both mental and physical, its intimate involvement in the physiologic activities of the brain, of the ear, and of the organs of the chest, throat, mouth and nose, its subconscious mental control, and the early period of its development, all contrive to afford many opportunities for a slip in the attainment of a completely successful developmental outcome. Once the function has attained to normal activity the breaking of that normality of action is relatively uncommon, and results only from extraordinary physical or mental disturbances.

Certain congenital anatomic or physiologic impairments of the brain, or of the peripheral organs involved, necessarily prevent the attainment of normal speech. And injury to any organ concerned in the speech function may necessarily have a similar injurious effect on the speech. Most of the disorders of speech of childhood are, as has been said, either congenital or developmental in origin, and such disorders, if not corrected, become incorporated into the settled speech as permanent defects lasting into the adult years, and often throughout life.

Whether the child fail to develop speech at all, whether the beginnings of speech development are markedly delayed, or whether, having entered on speech development, inability to perfect the function, be the difficulty, or whether in the midst of normal processes of speech development, a disturbance of the function be superimposed, the necessary relationship of the speech to the organs involved in speech, ear, brain, nervous system, chest, mouth and throat organs, is clearly necessary for their understanding. Therefore, not only are disturbances of functions of these organs capable of throwing light on the etiology of any disturbance of speech, but the reverse is also true, and disturbances of speech are capable of casting light on mental as well as other disturbances of function in the child. Thus speech disorders should take their place as a helpful factor in solving certain of the clinical problems of childhood.

In the normal child, as later confirmed and continued in the adult, an atmosphere of speech develops and permeates the mental life. Hardly any knowledge is acquired through the growing powers of perception by the growing action of the various sense organs, sight, smell, taste, hearing, feeling, etc., which is not acquired in conjunction with the hearing of words representing the knowledge acquired. While the child is learning the significance of the object, chair, its peculiar form, solidity, color, uses, etc., he is hearing the words "chair," "sit down," "rest," "hard," "rocker," "straight," etc., all of which ideas and words become gradually interwoven intimately into the accumulative significance of the child's idea of chair. In the mind this acquirement of knowledge with the acquirement of an understanding of words is represented by the intimate interrelationship between the various centers for perception with each other and with the perceptive center for spoken words. Thus the perception of definite word sounds, like "chair," occurs in immediate association with the words characterizing that object as "sit," and with the perceptive facts concerning the object, as "straight," "rocking," etc. Not only that, but the very associated acquirement of these words and facts, encouraged by the sentences heard regarding them, suggests immediately the putting together of words into sentences to express the knowledge as it is acquired. Not only a knowledge of the word and its meaning, but also a knowledge of sentence building is thus a natural accompaniment of the acquirement of a knowledge of the environment of the individual child.

Speech also is one of the most important natural means for the development of the physiologic possibilities of the mind. Living, itself, with the necessity of coming to understand the world, and to adapt one's self to the facts of life, constitutes the basal natural means for mental training. But speech carries this acquirement of understanding further. Not only does speech aid the childish mind to develop by feeding it on the explanations and reasonings of others, but the exercise of the child's mind as it seeks from others through speech a solution of its mental problems and in general expresses its own thoughts, constitutes a means of mental training of first importance. Not the least valuable feature of this mental exercise is the need for clearness of thought required by speech, and as the capability of the mind grows the development of terms representing generalizations and thought abstractions constitute actually vital "tools" for building the most perfect mental structure possible. Neither of the two other forms of language—gesture and written speech—possess nearly so important capabilities for mental training as does speech. So it is that the child who, like the deaf and dumb, may not acquire speech early in life, is seriously mentally handicapped. And the child who for other reasons than deafness may not acquire speech until unusually late is thereby, often to a serious degree, mentally retarded.

In approaching the developmental processes for speech, it is well to conceive of the child just born as made up of complex but undeveloped anatomico-physiologic potentialities, having reference on the one hand to the brain and nervous system, and, for our purpose, on the other, to the organic entities of chest, larynx, mouth and nose with their complex potentialities for muscular action. In the peripheral organs, action begins at once at the moment of birth, not, to be sure, in actual speech, but in movements closely similar to the movements later concerned with speech. The breathing, the crying and the swallowing are at first all reflex acts, which, however, involves muscle movements of importance later in talking.

At the same time the child begins its mental activities. These are instigated through the action of the sense organs, and all these organs, as has been seen, functionate in immediate relationship to that for hearing. The auditory word center thus begins to acquire impressions of spoken words almost at once after birth, these impressions being coincident with the acquirement of their meanings. The memory functions that hold in the mind both the word and its meaning develop little by little, until when the period of general mental development that determines the child's capability to essay attempts at talking has arrived, the auditory word memory is already stored, in a measure, for these beginnings. And as the talking goes on the motor (kinesthetic) word center, the center which holds in memory the sensations that serve to direct the complex peripheral acts of speech, also begins its development and education.

Crying is later accompanied by the production of many simple childish sounds, like "bah, bah, tah, tah," etc. These represent the natural expression of pleasure in muscular action, and later also a pride, as well as a pleasure, in the production of its own voice sounds. Still later imitation of the child's own production of sounds becomes evident. When at last the child attempts to repeat a word spoken by another person, it is close to the point where appreciation of the significance of speech is to begin. The mind has been steadily growing. Ideas have become more clearly apprehended. The auditory word center has become charged with crude memories of words and their meanings. The simple repetition of spoken words regardless of the meaning of those words is now supplanted by a higher mental process. The child by employment of its own speech organs, and with the purpose of expressing ideas, repeats words taken from its own auditory word center. The child has begun to talk.

From now on more definite education of the motor word center proceeds, just in proportion as peripheral efforts at speech production become more exact. Endless efforts and repetitions enlarges step by step the range of the peripheral sounds produced, and renders the effects more and more true. At first, speech is made up of "one word sentences;" simple combinations follow, and, finally, as the mind becomes equal to the more complicated effort called for, sentences become the natural manner of speech. The difficulties, both mental and peripheral, involved in this completed process may best be realized by recalling the years of effort required for their perfect elaboration.

THE FUNCTION OF SPEECH ON THE PERIPHERAL SIDE

If we bear in mind that voice (the basal sound of speech), like all sound, is dependent on vibrations of the air, and that the production of air vibration is a physical act, we at once realize the mechanico-physical character of the peripheral acts of speech. An understanding of speech and speech disorders requires that the conception of the peripheral apparatus of speech as dependent on an actual physical machine should be emphasized.

The requirements of the speech machine consists in the production of basal voice, of its modifications with respect to loudness, to pitch and to quality, and its maintainance of action under the complex and rapid changes of position required by the articulative necessities. The machine is called upon to modify the basal voice into (in the English language) some 45 definite elemental sounds, and to run these elemental sounds one following another, rapidly, or slowly, through almost every conceivable combination. To accomplish these ends the most complex musculature and the most complexly movable function in the body is required.

To dovetail so complex a function into the more fundamental life depending functions of breathing and swallowing constitutes one of the ingenius master strokes of the Divine Mechanician. The employment of the lungs as the fundamental store house of air for use in talking, as well as the chest wall for controlling for speech purposes of the accumulation of air in the lungs, was but the natural utilization of the more basal function of breathing. And the insertion into the tube employed to carry away the outgoing air for breathing, of a mechanism capable of setting that outwardly moving air into voice vibrations, although ingenious, was also relatively simple and natural. But the intricate interweaving into the apparatus required for swallowing of an incomprehensively complex musculature, in order to enable jaw, tongue, lips and soft palate to coördinate in a multitude of ways, both with one another, and especially with the extrinsic and intrinsic movements of the larynx, is an accomplishment too intricate for the present complete understanding of students of the physiology of speech.

The peripheral speech function is best understood by analyzing it into (1) the chest mechanism and (2*a*) the laryngo—(2*b*) articulative mechanism.

The chest mechanism of speech is nothing more nor less than the mechanism of breathing, but employed with the requirements of muscular effort, of irregularity, and of control demanded by the speech function. The principle of action is that of the bellows, but with complications of the bellows action resulting from the flexible diaphragmatic boundary between the chest and abdominal cavities. The expansive capacity of the chest wall and of the contained lungs, as well as the compressive action of the chest wall, while subject both to physiologic and to pathologic variation are practically never incapable of being adequate for an ordinary degree of voice production, so long as they are sufficient for breathing. For ordinary production of the voice requires only light muscular effort. On the other hand, however, marked developmental or pathologic impairment of the chest possibilities for expansion and compression may constitute an important hindrance to complete development of the vocal function. Both under the direction of Nature herself and under the direction of the voice trainer, the expansive and compressive functions of the chest are capable of important development both as to capacity and method.

The vocal cords which lie as a movable barrier in the path of the expelled air from the lungs, employ three primarily significant interrelated positions of approximation. First, so far apart as to be incapable of vibration (for breathing); second, approximated but with a narrow clear open intervening space and incapable of vibration beyond that required for the whispered voice; and, third, in alignment one opposite the other with a minute intervening space and capable of complete vibration. The first is the position for noiseless breathing, the second for the whisper, and the third for the loud voice. "Voice" technically refers to the vocal result obtained from the action of the vocal cords when in complete vibration. "Voiceless" refers to the vocal result when the vocal cords are in the whisper position.

The production of a "voiced" or of a "voiceless" sound is the result of muscular action of remarkable complexity. The larynx is not a fixed but is a suspended organ. It is not only held loosely and supported at the sides by the constrictor muscles and the fascia which holds it to the skin and to the esophagus and spinal column, but it is suspended from above by muscles which make it essentially a part of the musculature of the lower jaw and the base of the tongue, and by muscles which suspend it from the base of the skull and spinal column. From below it is supported and steadied by the hyoid and thyroid muscles that pass downwards to the sternum. The larynx, then, is, as a suspended body, subject to complex though limited movements in all directions, but especially obliquely up and down. Besides this the three cartilages forming the body of the larynx move with respect to one another.

For the production of any vocal cord activity, even of the simplest character, all of these four muscular systems, intrinsic laryngeal, inter-cartilaginous, upper extrinsic and lower extrinsic, must enter into definite activity, dependent on the character of the sound and on its pitch. Voiced sounds require strong fixation action of the extrinsic musculature, and voiceless sounds require much less strong fixationaction.

The general belief that the articulative function is a mechanism quite distinct from the laryngeal function is wholly incorrect. The basis of the articulative function lies in vibratory action of the vocal cords peculiar to each elemental sound, and dependent on a definite position of the larynx as a body, of the cartilages of the larynx, and on a peculiar action of the vocal cords themselves. In other words, the so-called articulative position upon which the especial elemental sound depends includes the mouth organs and the entire laryngeal mechanism. The sound result is dependent on the placement of this entire articulative-laryngeal musculature. A few sounds (like b and m, d and n, and g and ng, for example) seem to have essentially the same laryngeal action, but for the most part this action is distinctive for each elemental sound.

The production of the various individual articulative sounds (while dependent basally, as has just been said, on the action of both the intrinsic and extrinsic laryngeal musculature) is dependent also on the principle of sound resonance as determined by size and shape of the mouth and adjacent cavities; on the principle of partial retardation of the expulsion of air ("friction sounds," f, v, th′, th″, s, z, etc.); and on the principle of momentary complete stoppage of the outwardly moving air current ("explosive sounds," p, b, t, d, k, g).

The resonantial value is especially important for the nasal sounds. The soft palate is capable of complete closure of the oro-naso-pharyngeal aperture, or of partial closure, or of no closure at all. The effect on the elemental sound depends on the degree of access of the laryngeally vibrating air to the nasal spaces. For the nasal sounds this opening into the nose is complete. A careless closure of the palatal mechanism results in a variably nasal voice.

Classification of the elemental sounds should have reference to the mechanics of the production of the individual sounds. The following classification is modified from that long employed by Dr. G. Hudson Makuen, and was by him obtained, I believe, largely from Dr. John Wyllie.*

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* Wyllie, John, M.D., "The disorders of speech." Edinburg, Oliver and Boyd (1894).

VOWELS

ē	ā	ah	aw	ō	o͞o
even	apes	are	awed	over	oozing
ĭ	ĕ	ă	o͝o	ŭ	ŏ
if	Edward	adds	books	up	on

VOWELS

ē-o͞o = ū, use
ah-ē = ī, ice
ah-o͞o = ou, out
aw-ē = oi, oil

ä ask, last
r her, burr
a-r air, care

CONSONANTS

	Voiceless Oral	Voiced Oral	Voiced Nasal	
Labial sounds	p	b	m	Paul, Brown, made
	wh	b	..	white, wax
Labio-dental sounds	f	v	..	full, voice
Linguo-dental sounds	th′	th″	..	think, thou
Anterior	s	z	..	some, zealous
Linguo	.....	y	..	yearling
Palatal	sh	zh	..	sheep leisurely
	t	d	n	took, down
Sounds	.....	....	..	nine large
	.....	l	..	rails
	.....	r		
(Compound)	t-sh	d-zh	..	cheap John
Posterior linguo-palatal	k	g	ng	Can girls sing
Multiple position sounds	h	....	..	heap, hope

Addenda	Voiceless Oral	Voiced Oral	Voices Nasal	
X	k-s	....	..	axe
	...	g-z	..	example
Qu	k	....	..	oblique
	k-w	....	..	quick
C	s	....	..	city
	k	....	..	cooks

THE SPEECH DEVELOPMENT PERIOD

Nature has set no arbitrary moment when the child shall be ready to enter on the development of speech. The period of preparation for the onset of this peculiarly human function is concerned with the beginnings of mental development, with the learning to appreciate speech sounds, and with the preliminary exercising of the peripheral speech organs. Sometime between the end of the first year of life and the end of the third year, the child essays the attempt to repeat spoken words; but the time of onset, to be normal is but relatively fixed at either end of this period. Children who begin to talk at the end of the third year may progress to the acquirement of normal speech, and may be found to possesss intellectuality quite as normal as that of the child beginning to talk in the second year. But, nevertheless, the children who, later, show deficiencies in their capabilities for acquiring completely normal speech, or for full capability of intellectual development, are apt to have begun to talk relatively late. Whether, then, the continued delay of a particular child to start talking as the third year goes on is, or is not, significant of some manner of deficiency in the child may well become in such children a matter for determination through study of the various characteristics of the child. Still another consideration of importance in this connection has reference to the long period of time ordinarily required by the child for the full development of speech and to the fact that as this length of time tends to come to an end—at the seventh or eighth year—the natural inclination and capability of the child to concentrate on the speech and to labor spontaneously for speech correction has begun to diminish. Thus a relatively early inception of the speech—but not phenomenally early—is not without its importance. Moreover, the earlier the child begins to talk the sooner are the intellectual developmental uses of the speech entered on. Roughly speaking, then, the onset of speech should not be considered pathologic before the end of the third year of life, and the "speech development period" may be said to comprise the period between the ends of the first year and the eighth years of life.

Children who begin to talk after the end of the third year may also, to be sure, go on to normal speech and may prove to possess normal intelligence; but, this longer delay of the onset of speech is likely to point to a physiologic inaptitude for speech development of some sort, or to an actual pathologic hindrance to the acquirement of speech. If the child begins to talk very late, and, although capable of complete speech development spontaneously under normal conditions of onset, the development of speech, because of slowness in the process, has not been completed at the end of "the speech development period," imperfections of the articulative processes may continue as fixed imperfections, and become settled habits. Such habits imply the permanent establishment of memories in the auditory word center, the incorrectness of which the individual has ceased to be cognizant of. Such fixed cerebral errors in the sensory memory processes of speech are continued into the motor word memories, and become fixed in the movement processes of the peripheral articulative organs. This cessation of spontaneous effort to improve incorrect articulative processes occurs with children of normal intellect, and of course more especially with children whose intellect, or other faculties concerned with speech development, are impaired. The phenomenon serves to emphasize the importance of the conception of a normal speech development period, and should call one's attention to the dangers of too great slowness in the speech developmental processes.

MUTISM

Classification and General Etiology

Talking expresses a deeply human aspect of the physiologic nature of the child. Mutism, therefore, signifies a serious reversal of one of the child's most important birth rights. Given normal anatomical foundations, normal health conditions and normal environmental conditions, and the development of speech in the child should follow as certainly as development in height and weight and general intelligence. But the acquirement of speech is a complex procedure involving united coöperation between different organs and physiologic processes, and, therefore, is subject to various possible upsets.

The machinery of spoken language is made up of all the peripheral organs of sense (in coöperation with that for sound), of the brain with its various centers, of the peripheral organs of speech, of the connecting nerve tracts for these various centers and organs, and of the concerned muscles. The forces at work are those of peripheral sense perception (more particularly for sound), and of mental, nerve and muscular activity. Acquirement of normal speech involves the necessity for essential normality in all these various organs and physiologic forces. Through their normality of action all these organs and forces gradually learn to work in harmony and with a high degree of skill for the attainment of their highly complex speech purposes. Through the peripheral organ of hearing enter the peculiar sounds required for speech, and through the ocular and other sense organs enter the ideas which give meanings for these sounds. By developmental processes of growth the general mental powers gradually become increased; and thus both the thought content for speech and the mental incentive and intelligence required for the employment of the peripheral speech organs in talking become gradually developed. And as talking is begun, the education of the motor word center proceeds.

Mutism implies some serious upset at some point in this elaborate physiologic complex. If the external ear be seriously impaired the nutriment of spoken sounds cannot feed the auditory word center, and the mind then possesses no sound-word material on which to build the speech. If the auditory word center itself fails to functionate the same effects result. If the general mental development fails, the mental content for speech, i.e., ideas, is to a greater or less extent lacking; and, besides, the mental forces on which depend the education of the sensory word center and the incentive to employ the peripheral organs of speech for talking, are both either completely or incompletely absent. One may also be compelled to seek in the peripheral organs of speech, or in the motor word center, or in the connecting nerve structures, for the explanation of mutism.

For the convenience of clinical study we may classify mutism broadly as follows:

(*a*) *Permanent Mutism.*—No possibility of ever acquiring speech either through natural or artificial means.

(*b*) *Deaf Mutism.*—Mutism permanent unless acquired through direct artificial education of the peripheral speech organs, and thus, indirectly, the motor-word center.

(*c*) *Temporary Mutism.*—Beginnings of speech delayed beyond the usual period for onset, but the speech capable of development either perfectly or imperfectly; at any rate with, if not without, especial aid.

Approaching the subject from the clinical standpoint one should realize that the term mutism must of necessity, be employed with reference to the life period of the child, and with reference to its transient, or permanent, possibilities. The normal child who at two years does not yet talk is not mute in any but a normal sense. But the boy, or girl, who at five years does not talk is at that time pathologically mute, even though speech may be entered upon later. Unphysiologic mutism (from a time standpoint) begins after the normal age for the beginning of speech has passed. This limit for the ending of the period for normal spontaneous development of speech we may place roughly at the end of the third year, after which the unspeaking child may be said to be pathologically mute, whether the mutism be permanent for life, or temporary for any period of years.

Whatever be the immediate cause of mutism, questions of mentality must ever insinuate themselves into the involved etiologic and therapeutic problems. For, learning to talk is not only a means for thinking aloud but it is of itself dependent on a certain degree of normality and of virility of mental action for the development of its own complex processes. A normal, vigorous mind is much more likely to overcome handicaps to speech development than is a partially normal and sluggish mentality. Abnormalities along the speech tracts, if they be not themselves vitally important to the speech, derive their significance in large part from their possible relation to a vigorous, or a non-vigorous, general mentality. Relatively slight localized impairment, for example moderate hardness of hearing, or the presence of adenoids, or moderate paresis of the tongue, might be causes for mutism in a child of impaired general mentality, but might have little retarding influence in a child of virile mind. Also if congenital, or pathologic processes after birth, result in impairment of the general mentality uniformly, including the speech tract itself, to a degree still admitting of the development of speech, then speech may usually be expected to result; but if the central speech tract happens to be more profoundly incapacitated than the general mental areas mutism may be the consequence. Again, environmental handicaps such as shall be mentioned under temporary mutism are of importance largely in proportion to the mental normality of the child.

In children who are mute because of peripheral defects, especially the deaf mute, it is necessary to realize that defects of mentality may also be present. The feebleminded deaf mute must be classified differently from the normally minded deaf mute. For the former is quite incapable of acquiring instruction which the latter may acquire, and is entirely out of place in an institution for the deaf. But it is also to be realized that the long uninstructed normally minded deaf mute is usually but little capable of mental analysis and abstract thinking, and if we are not on our guard may be wrongly and injuriously classified with the feebleminded.

General Diagnosis of Mutism

The diagnostic problems presented by the mute child are various and often puzzling, and may require much experience and much study. The diagnosis is always linked up closely with prognosis: Is the mutism temporary or permanent? The anxiety of the parents to know whether the child can ever talk is not to be put aside. Early diagnosis is important, in order that the precious years of "the speech development period" may be so far as possible utilized for the complete development of speech, where speech can be developed. The diagnostic approach to the problem takes into consideration: (1) The peripheral hearing organs; (2) the mental status; (3) the peripheral speech apparatus; (4) environmental considerations.

The diagnostic approach to the mute child is by way of the peripheral organ of hearing. If the child is so hard of hearing as to preclude the understanding of spoken words, the immediate cause of the mutism is explained. If the mentality in such a child is evidently normal and the deafness sufficient to prevent the development of speech, the child is classified as a deaf mute of the ordinary type. Such cases present no especial diagnostic difficulty.

But if the child be deaf to spoken words, and in addition presents mental peculiarities which open a doubt as to the existence of a low degree of feeblemindedness, the problem becomes more complicated. To be sure the deafness determines the immediate cause of the mutism; but the manner of future treatment of the child is dependent on the still undetermined question of feeblemindedness. The latter type of child is quite incapable of receiving the ordinary instruction for the deaf and dumb and finds no place in ordinary deaf and dumb classes or institutions. On the other hand, the deaf mute child of essentially normal mind may present mental deficiencies and peculiarities of manner, which place his essential mental normality in doubt. The long uninstructed deaf mute is more or less seriously defective in knowledge and reasoning power, and, if unintelligently brought up, may, owing to mental isolation and ignorance of the common conventions of social relations, appear ignorant and peculiar enough to be mistakenly classified as feebleminded. Cases of this character are likely to require persistent and close observation, with the purpose of determining the mental capability and the teachability of the individual studied.

Another situation with respect to the mute child who is incapable of hearing spoken words is found when the child is seen to be frankly and seriously feebleminded. This picture is that of the deaf and dumb idiot or imbecile. The combination is not common. No question of the seriousness of the mental condition is likely to arise, but owing to the absence of capability of attention considerable careful observation may be required to settle the question of deafness. From a practical standpoint, however, if the high degree of mental impairment precludes the possibility of instruction, it does not matter whether the mutism be primarily due to central or to peripheral causes.

In the mute child a study of the hearing, which constitutes, as has been said, the most natural approach to the diagnostic problem, may disclose a degree of hardness of hearing which permits of an appreciation of speech only when spoken with much greater loudness than that of ordinary conversation. Under those circumstances the apprehension of speech is filled with uncertainty at every point, the appreciation of the sounds and words of speech is crude and uncertain, and the memories of the auditory word center are of the vaguest character. To realize the relationship of the hardness of hearing to the mutism is here one of the diagnostic problems. If the mentality is normal and especially if mentally bright, the child would be likely to create the best articulative procedure of which his defective hearing made him capable, and attempt to make up for the lack of understanding of speech due to the hardness of hearing, by an instinctive effort at lip reading. But if the mind be sluggish and especially if the child is feebleminded, such a handicap might be the immediate cause of the mutism. In this connection it is to be noted that hardness of hearing is common in the feebleminded. Owing to the feebleness of the mental energy and to the lack in mental clearness, to the absence of understanding of the facts of life, defective hearing even of moderate degree may prove an insurmountable obstacle to the acquirement of speech in children so afflicted.

Beginning now to turn to the cases of mutism in which the peripheral organ of hearing functionates normally, two conditions with respect to the understanding of speech may be present: Either (*a*) the child understands and responds by action to what is spoken, or (*b*) he does not understand, and consequently does not respond by action. In the first instance the cause of the mutism must lie not only beyond the peripheral organ of hearing, but also beyond the perceptive word function. In the second case the perceptive word function is not operating normally, and is presumably the cause of the mutism. The latter are the cases of "psychic deafness." The hearing is capable for sounds of ordinary loudness, and yet understanding of speech meanings is absent. Such children sometimes prove to be of nearly normal intellect, although serious degrees of mental impairment are more common. Appreciation for music may be present, and they may be taught to sing wordless tunes. Repetition of spoken words, but without appreciation of their meanings, may be possible. The mental deficiency is not in the appreciation of sounds as such, but is in the apperception of the meanings of words. Whether the cause for this central deficiency lies in a specific failure in the development of the "apperceptive word center," or in the memory function of that center, or whether the deficiency lies rather in a lack in the functions of attention and concentration must at this time be considered unsettled questions.

If the peripheral organs of hearing be essentially normal, and the apperception of the meanings of spoken words be present, the explanation of the mutism must lie in the mentality beyond, so to speak, the perceptive word center, or in the peripheral organs of speech, or in both. Developmental abnormalities of the peripheral apparatus—teeth, tongue (tongue tie), palate, naso-pharynx (adenoids), or even a considerable degree of paresis, do not, unless possibly with rare exception, produce mutism if the mind be entirely normal. The incentive to speech is so instinctive and fundamental in the child of normal mind that any but the severest handicaps are likely to be overcome to the degree that the organic impairments present permit. Therefore mutism in the child of impaired peripheral organs of speech calls for study of the mentality.

The psychologic incentives to speech are primarily imitative, and emotional rather than intellectual. The child experiences a desire, as for drink, and calls for "water;" or he experiences the sensation of burning and exclaims, "hot!" The process of intellectuality enters but lightly, and is required only to a very simple degree for the simplest one-word speech. Intellectuality of some degree is required for a considerable one-word speech, and to a much greater degree for a speech involving the employment of sentences. One expects, then, the development of a few words of speech in the idiot or imbecile whose hearing is normal, and expects a more complex speech in proportion as the degree of mental impairment is lessened, even to a fairly fluent speech and a fairly large vocabulary. Such expectations may, however, be completely unfulfilled for reasons not definitely explainable, but usually in all probability related to some localized impairment in the speech tract of greater severity than that involving the brain as a whole.

The determination of the mental capability of the child who is mute is naturally more difficult than for the child who talks. This natural greater difficulty is emphasized when one realizes that the scheme of testing for mental capability devised by Binet and Simon is based very largely on speaking capabitily. The more recently developed performance tests, as exemplified by those of Pintner and Patterson, aid materially in solving the mental diagnostic problems of the mute child.

PERMANENT MUTISM

The impulse to talk is virile in the normal young child. In the course, therefore, of the early years of life discouragements other than those of decided physiologic significance are very likely, sooner or later during this early period, to be in whole or in part overcome, and speech in some form spontaneously to break through. The children who can hear and yet who can never hope to talk are, therefore, in some manner profoundly unphysiologic with reference to speech. They are children (1) with profound general mental retardation (idiocy, imbecility); or (2) children who are generally mentally retarded, though perhaps to a less degree, but in whom apparently some more profound impairment in the mental speech territory has occurred than that which exists in the general mentality. Or (3) children who through maldevelopment, or accident, or disease, are profoundly paralyzed or otherwise handicapped with respect to the peripheral speech region.

The condition of mutism in these highly abnormal children tends to merge imperceptibly into that of a primitive capacity for speech, and no accurate line can be drawn between the conditions of complete speechlessness and of primitive speech. And, indeed, no line can be drawn, excepting arbitrarily, between the condition involving the simplest speech and that of the more elaborate but still abnormal speech. The mute child, as the term is used in this book with reference to the feeble-minded, includes only conditions in which the vocabulary is limited roughly and arbitrarily to a half dozen or dozen words, or less, crudely enunciated, and where sentence building is absent.

The extremely low-grade idiot may be essentially lacking in any manner of self-expression. Incentive to speech lies in the expression of feeling and other emotion, as, for example, fear, or desire, rather than in the expression of thought, which indeed may be absent, or else too indefinite for expression. Children of extremely low mentality may never develop self-expression beyond a few simple gestures; they may make crude and meaningless animal cries; or they may attain to a few hardly understandable monosyllabic words, which they employ in one word sentences. As the scale of general intelligence rises the vocabulary may also rise.

These children usually present themselves as evidently idiotic or imbecilic. While no actual physical stigmata of degeneration need necessarily be present, the frequent impairment of movement in walking, or the drooling, coupled with the history, and other evident indications of marked mental impairment usually make the situation clear to the experienced physician. Two well-defined temperamental types present themselves, the apathetic and the active, with all degrees less clearly defined in between. The extremely apathetic child may sit staring blankly for an indefinite time, showing no interest in its surroundings. The active child may never remain still, but keep up an endless, almost purposeless movement, showing only momentary interest and attention to its immediate surroundings. Effort to secure the attention of individuals of either type, even for an instant, is apt to be unsuccessful.

Another group is made up of *the psychically deaf*. The mentality varies but may in certain cases upon treatment approach near to normal. Because of the hopeful possibilities of treatment in certain of these cases it is important that they should be correctly classified. Sounds of ordinary loudness can be heard, and yet spoken words cannot be understood, and in spite of the fact that gesturing and the conduct generally indicates an intelligence that should be sufficient for the understanding of speech. Imitation of spoken words may be possible but without appreciation of their meanings. Appreciation of music is present. That part of the mind which coördinates the sounds of words with their meanings as elicited by other sense organs is the part immediately at fault. This function requires undoubtedly a certain degree of attention and concentration and even of analysis. If the hearing can be shown to be present and the mentality and capability of attention to be considerable, and yet understanding for speech to be absent, the case is quite clear. The lower the mentality the more difficult the exact diagnosis of both the hearing and the mental capability. They are likely to be mistaken for deaf mutes. Differentiation depends on the proof of their hearing of sounds of low intensity, for they understand speech no better than deaf mutes. The differentiation of this class of mutes from the simple feeble minded children who are generally incapable mentally of holding the attention closely enough to understand speech, lies in the very fact that the mentality of the psychically deaf seems to admit of the capability of attention and concentration required for speech. H. Gutzmann finds the explanation of the failure to appreciate spoken words in the failure of the memory function of the perceptive word center, and T. Heller thinks that in some cases there may be a specific failure in the capability of the attention. Close and persistent study is required for clear diagnosis, which, since not all of this group are incapable under treatment of acquiring speech, is of especial importance.

The child who has acquired speech, but who through disease or accident possesses an incapacitated sensory word center (sensory aphasia), is likewise both psychically deaf and also capable of appreciating sounds of ordinary loudness. But such a child may retain some capability of speech, and the history of previous understanding and production of speech will help to settle the etiology. The mutism in some of these children will also prove to be only temporary.

The child who is paralyzed in tongue, or lips, or larynx, is also likely to show indications of impaired mind, presumably due to the same cause that produced the paralysis. The causes may be meningitis, birth injury, syphilis, falls on the head, etc. If the mind be quite clear the child will usually make some manner of attempt to talk. If the mind be sluggish the mental energy demanded to overcome the handicap of paralysis is lacking, at least unless through careful training. If the speech be in some manner developed it is necessarily inexact, thick, and more or less difficult of understanding. This depends on the degree of paralysis and the degree of mental impairment. If the paralysis exhibits the characteristics of spasticity the problem is more severe. Even though the mentality admits of the development of the speech the production of spasm by each attempt to talk serves to render the training necessary to the development of speech too wearing to be practicable. Here, however, also the question of clearness of mentality and intensity and extent of the paresis and the severity of the spastic tendency, all have their bearing on the possibility of developing speech.

DEAF MUTISM

The peculiar significance of deafness in the young child, as compared with the adult, is dependent on the relationship of the function of hearing to the acquirement of speech. If the child cannot hear, the basis for normal speech development, the development of the auditory word center, is effectually prevented, and thus both the understanding and the reproduction of speech in the usual manner become impossible. The deafness of the deaf mute must either be present at birth, or must be acquired prior to a certain life period of the child; and it must be of a degree sufficient to prevent the education of the auditory word center in the ordinary manner. Such a degree of deafness has been specifically named by J. Kerr Love as "surdism." The child is deprived not only of the educational value of the speech of his environment, but also of the great mental developmental value attendant on the learning and employment of spoken language. He becomes individually and socially an especial and serious developmental problem.

Closely allied, but not classifiable directly with surdism, is a degree of hardness of hearing in the child, which, while severe enough to greatly interfere with the development of speech through the agency of the ear, is still insufficient, at any rate with expert aid, to prevent such development. The natural channel for the acquirement of speech is so important and so superior to other artificial channels that the distinction between surdism and hardness of hearing must ever, from an educational standpoint, be kept clearly in mind. In the days gone by it was not uncommon to educate seriously hard of hearing children as if they were deaf mutes, and, while this mistaken procedure has undoubtedly become much less common, one is suspicious of its too frequent occurrence even today.

The child afflicted with congenital surdism is as a matter of course a deaf mute, for practically none recover their hearing sufficiently to lift them out of this class. But approximately half of the whole number of deaf mutes are not congenitally deaf. Some acquire deafness in the first year or two, before speech has well begun (a very large proportion of the acquired cases), and some after the second year, either when speech has been but crudely begun, or even when it has become quite well established. Whether the advent of deafness in a speaking child shall eventuate in deaf mutism depends on such factors as age, the degree of proficiency in talking, the mentality, the general health after the deafness has developed, and the promptness and energy and efficiency with which measures to retain the speech are undertaken. Nadoleczny points out also that knowledge of writing is an aid to the retention of the speech. Previous to the seventh or eighth year speech is seldom retained, unless with the most careful aid, although children as young as four have, with guidance, retained their speech. Other children as old as 12 or even 14 have become deaf mutes.

The deafness of the deaf mute is the result in all, or nearly all cases, of impairment not of the middle but of the internal division of the ear. Hardness of hearing, on the other hand, is typically the result of middle-ear impairment. Middle-ear disease alone does not result, unless possibly very exceptionally, in a degree of deafness so extreme as surdism. Bony occlusion of the internal auditory canal (a rare congenital defect), however, or occlusion of both the oval and oblong windows separating the middle ear from the labyrinth produces surdism. Whether the deafness be congenital, or acquired, it is, then, almost, if not always, the labyrinthine structures that are at fault. Infections of the labyrinth may find entrance either by way of the middle ear, in which case the middle ear, including the drum-membrane would usually show indications of past disease, or by way of the brain, in which case the middle ear, including the drum-membrane, would, unless middle-ear disease had supervened, remain intact. Infection of the middle ear may progress into the labyrinth, chiefly by way of the fenestræ. And infection from the brain may find its entrance either along the auditory nerve or the perilymphatic or vascular channels of the nerve structures.

The pathologic anatomy of deaf mutism has not received the extensive study that its importance calls for. In 1912 Dr. G. Hudson Makuen stated that not a single report of post-mortem study of a deaf mute could at that time be found in the medical literature either of Great Britain or the United States. So far as the author knows, nothing of importance in this direction has been added in these countries since that time. Fortunately, however, through the work of Holger Mygind, Bezold, Politzer, and others, 150 to 200 post-mortem studies of deaf mutes have been made. In spite of the importance of these studies in throwing light on the pathology of deafness in the deaf mute, Doctor Makuen calls attention to the relative lack of value in these studies, because of the failure to connect each case studied with its history and clinical manifestations. Such facts as the clinical status of the hearing, the existence of other deaf mutes in the family, the disease producing the deafness, the mentality, the effects of treatment, taken in connection with the pathologic findings, would tend to throw light on the physiology of the hearing organ, the pathology of heredity in the deaf mute, the local pathology of diseases that produce deafness, methods of treatment, etc.

Inflammatory processes occur in both the middle and internal ear, as well in fetal life as after birth, and the pathologic findings apparently do not as yet furnish a basis for distinguishing between these processes. The pathologic findings include atresia of the external auditory canal (in both congenital and acquired cases), obliteration of the tympanum, and partial or complete bony obliteration of the round and oval windows. And in the internal ear a great variety of pathologic findings are found. Malformation, or failure of development of the membranous labyrinth, or of the auditory nerve, are met with in congenital cases. The semi-circular canals are most often affected, then the cochlea and finally the vestibule. The changes involve degeneration of the membranous structures and then replacement with fibrous, calcareous, or bony substance. In the cochlea, the organ of Corti, the tectorial and Reisner's membrane may be injured or destroyed. The auditory nerve and its branches at times show degeneration. In general the pathologic findings are not known to be different in kind, although perhaps more severe in degree, from those producing a corresponding degree of deafness in adult life.

The older classification of deaf mutes, according to the time of the acquirement of the deafness, i.e., into congenital and acquired deaf mutism, fails, as Dr. J. Kerr Love points out, to satisfy the needs either of the student of etiology or of prevention. The more specific classification of this author opens the way to clearer analyzation of the causes of deafness, and paves the way for a still more specific classification as our knowledge of causes grows. The classification of Doctor Love is:

I. Deafness undoubtedly acquired after birth.

II. Sporadic Congenital Deafness.—Some of these cases may have been acquired during the first year of life, or even as late as the second year.

III. True Hereditary Deafness, as respects children always congenital; its distinctive feature is that the family history always shows other cases of deafness, either in the direct line or in collateral branches of the family.

To prove whether deafness in the child is congenital, or acquired, is not always possible. The deafness may not be perceived before the second or third year. Then, even though the child may have had a serious illness previous to this discovery, like meningitis, or scarlet fever, or may possibly have had a fall, proof that the deafness did not exist previous to that instance of disease or accident may be impossible to obtain. These necessarily doubtful cases are classified by Doctor Love as sporadically congenital.

According to various authors (Bezold, a little more than one-half; Holger Mygind, about one-third; Love, about one-half) the acquired cases of deafness in deaf mutes number from one-third to one-half the total number. Doctor Love points out that the more accurate our statistics become the larger becomes the proportion of acquired cases. Of the acquired cases by far the most common cause of deafness is meningitis (including "cerebrospinal convulsions," "hydrocephalus," "congestion of the brain," syphilitic and tuberculous meningitis). The next most common disease, scarlet fever, may produce deafness by first producing meningitis, or by infection of the inner ear after first invading the middle ear. The same is true of other infectious diseases. If the infection is limited to the middle ear, any resulting effect on the hearing is indicated by hardness of hearing rather than "surdism." Other causes producing deafness and consequent acquired deaf mutism, entered somewhat in the order of their relative frequency, are, purely local suppurations of the ear, traumatism, syphilis, measles, diphtheria, mumps, typhoid fever, pneumonia, osteomyelitis, whooping-cough, etc. The rarity of meningitis and scarlet fever as childhood diseases, when compared to measles, for example, serves to emphasize the importance of scarlet fever and especially of meningitis as causes of deaf mutism.

Syphilis stands alone (since otosclerosis does not produce deafness in childhood) as a disease which causes deafness after birth and consequent deaf mutism, in which entrance of infection into the child's body occurred back in prenatal life. The injury to the ear from syphilis may occur in the embryo, or in the first year or two (syphilitic meningitis), or later in childhood, or even in adult life.

Doctor Love, whose devoted and scholarly studies of the deaf mute form much of the basis for this portion of this article, has made clear the importance of differentiating between "true hereditary" and "sporadic congenital" deafness, and has laid the ground work for such a differentiation. The need for such differentiation has, however, been known for many years. The term "hereditary" should be applied only in cases in which the hereditary taint was present at the very moment of conception, Doctor Love argues, and should not be applied to pathologic taints that have been acquired in embryo. The life of the child in this connection begins at the moment when the male and female elements unite to form the ovum; one or both of which elements lacks some manner of potentiality required for the creation of a perfect child. All other cases he calls "sporadic congenital" deafness. His clinical differentiation between hereditary and sporadic congenital deafness is based on certain facts: First, both hardness of hearing and deafness result from many causes, and, therefore, even when occurring in the offspring of parents, cannot of themselves alone be adduced as evidence of a hereditary tendency. Second, syphilis stands alone as the one disease capable of causing hardness of hearing in the parent and of being transmitted to the child as deaf mutism; hardness of hearing in a parent of a deaf mute child should, therefore, never be adduced as evidence of heredity of deafness. Third, congenital deaf mutism occurring in brothers and sisters is (because of the possibility of congenital syphilis as a common cause of the deafness) not so good evidence of heredity as deaf mutism occurring in a collateral branch (cousins, uncles, etc.). Doctor Love narrows the evidence of heredity down to the finding of one or more deaf mute relatives among the progenitors, or collateral branches. A hereditary deaf mute is one, therefore, according to his definition, who was born deaf and who has had one or more deaf born relatives (especially if these were not the brothers or sisters of the deaf mute in question). Very rarely it happens that a child is born hard of hearing and not deaf. Such children, however, might possibly represent those really deaf children whose "islands of hearing" happen to stand in the pitch range of the speaking voice. If the latter were the fact cases of this class would satisfy Doctor Love's rules of evidence as to heredity of deafness. Moreover, cases of congenital deafness which are not known to possess congenitally deaf relatives might, if the family statistics were extensive enough, be found to have had such congenitally deaf members. Consequently one cannot rule out many of the congenitally deaf as evidence of family heredity, and also often cannot, of course, admit them as such evidence.

It is difficult to study any possible hereditary family taint statistically. First comes the difficulty presented by carelessness of history taking, and, second, the difficulty of tracing progenitors and relatives sufficiently remote to make the statistics adequately tell their story, and finally, the tabulation of statistics by different students is made on such different bases as to render comparison of statistics only partly possible. However, the monumental work of Dr. E. A. Fay made it clear with respect to marriages between two congenitally deaf mutes, that the percentage of marriages resulting in deaf offspring is high (28.4 per cent. of the marriages), and that the percentage of deaf children is likewise high (30.3 per cent. of the children). His statistics also show that marriages of deaf mutes in whom deafness was acquired results ordinarily in no deaf offspring. But, on the other hand, if hereditary deafness has existed in the families of those who have acquired deafness, a large percentage of the offspring are deaf. And, more than this, when hearing persons in whose families hereditary deafness is present marry, a large percentage of congenitally deaf children results. Doctor Fay's studies show also that the highest percentage of marriages of all resulting in deaf children (45.1 per cent.) occurred when congenitally deaf mute relatives intermarried, and that the percentage of deaf children (30 per cent.) was also high. These statistics have in all essentials been verified by later statistical students, including Doctor Love. This author, after careful analyzation of all the facts involved, concludes that nothing at present known precludes the possibility of the operation of the law of Mendel with respect to the propagation of hereditary deaf mutism. Proof of the operation of such law must rest on extensive statistical data running through a number of generations, and this information is not now fully existent.

Hereditarily, deaf families are of two classes. In one deafness alone is carried, and in the other deafness plus feeblemindedness, or epilepsy, etc. Doctor Love points out the fact that the degenerative tendencies of certain hereditary deaf mutes are not dependent on deafness, but are probably other manifestations of the underlying cause that is common to both the ear and other defective organs, like the brain. We shall see later that the hereditarily deaf mute is ordinarily a person of good general mentality. In accounting for these various defects that at times accompany the deafness, Doctor Love calls attention to the fact that the structures of the internal ear are derived from the embryonic epiblast and that the other structures showing defects in families carrying hereditary deafness are also derived from the epiblast. He points out that common hereditary factors involving the epiblast alone may explain from a hereditary standpoint all of these defects. It must not be understood, however, that heredity accounts for all the defects of mentality found among deaf mutes. Some of such cases belong among the sporadically congenitally deaf, and among the acquired deaf mutes.

Sporadic Congenital Deafness

Statistical figures adequate to determine the relative size of each group of the deaf mutes are not yet in existence. Such statistics as do exist may be generalized for practical purposes as follows:

Acquired deafness, 50 per cent.
Sporadic congenital deafness, 30 per cent.
Hereditary deafness, 20 per cent.

One should bear in mind that more accurate knowledge of the individual cases might compel the transference of certain ones from the acquired to the sporadic congenital group, and others from the latter to the hereditary group.

The origin of the deafness in the sporadic congenital group is less clear than for the other two groups. However, Doctor Love has shown that at any rate for Glasgow a very large majority of these children come out of the poorest and most crowded families. This means "a one apartment standard of living, of air supply, of food, of clothing, and it results in a dwarfed child;" and more than that, "it means untreated syphilitic disease, uncontrolled use of alcohol in many cases, a carelessness in the upbringing of children, and these, when added to the other conditions, result in an enormously high death-rate, and in a deaf mute rate which is never approached in the homes of the well-to-do."

The importance of congenital syphilis as a cause of deafness in children seems to be becoming more and more realized. The Wassermann reaction has helped to stimulate interest and study. For example: Ardenne obtained 13 positive Wassermann reactions in 23 deaf mutes examined. Doctor Love analyzed 21 undoubtedly syphilitic families and found that "nearly two-thirds of the children born are dead, or if they are alive are either deaf or blind or both (106 in 172), and with few exceptions there are no adults in the family. In many of the families one or more deaths from meningitis have occurred." Among the above 172 children, 31 were deaf, or deaf and blind. The blindness from syphilitic keratitis is a fairly common accompaniment of the syphilitic deafness. The meningitis of syphilis may occur before birth, often during the first or second year of life, and even beyond the thirtieth year. Death may be its outcome, or deafness, blindness, paralysis, or impaired intellect. Again referring to the 31 deaf children of syphilitic families discussed above, about 70 per cent. of the cases of deafness occurred after birth, many of them within the first two years of life.

The pathos of the situation lies in the fact that syphilis is a curable disease, and that these results in deafness could by adequate treatment be avoided. Doctor Love points out that syphilis also occurs in the families of the well-to-do, but that these terrible effects in the children are relatively very uncommon.

The manner in which alcoholism acts to produce congenital deafness, if it actually does so, is not exactly clear. Its action is coupled with the consequent neglect of the child in respect to all the conditions required for normal child development. And one may presume that the production of deafness is linked closely up with the production of epilepsy, and feeblemindedness, and sometimes with syphilis also acting. At any rate when we have clearly realized that the origin of sporadic congenital deafness is to be found in the consequences of poverty, crowding and neglect, we are started on the right road to a further understanding of these serious disorders.

For the most part deaf mutes are not totally deaf. Bezold found that one-third of the inmates of institutions possessed hearing sufficient to furnish aid in the acquirement of speech. Out of 138 persons tested he found only 27 (19.7 per cent.) totally deaf in both ears. Doctor Love and Doctor Rossie agree that: (1) Not more than 14 to 20 per cent. are deaf to aerial conduction, and a much smaller percentage to bone conduction; second, about 25 per cent. possess hearing of value in acquiring speech, and 10 to 15 per cent. are only semi-deaf; third, cranial conduction is almost always present. From these figures it becomes clear that deaf-mute institutions include not alone deaf mutes proper, but also those who belong rather in the classification of the hard of hearing. If a particular child possesses hearing sufficient, even with the help of artificial hearing aids, to permit of the complete education of the auditory word center, his outlook on life certainly should not be that of the deaf mute, but rather that of the hard of hearing. A further discussion of differences of hearing will be taken up under education.

The intelligence of the deaf mute is dependent on two factors, first, the normality of the intellectual structures themselves, and, second, the deterrent effect on intellectual development of the deafness. The child who has a purely localized impairment of the hearing organ is potentially as capable intellectually as the corresponding child who has no injury to the ear. However, in so far as the same causes that produced the injury to the ear, or other causes, may have impaired the brain structures also, naturally the intellectual capability must be correspondingly impaired. Undoubtedly the same conditions of impaired nourishment, of neglect, of prolonged ill health, etc., involved in the production of injury to the ear have also in certain cases produced impairment of the general physical and intellectual capability. This is true of certain of the sporadic congenital, as well as of the acquired cases. In meningitis the brain may have been left impaired; and is so also in hereditary cases combined with arrested brain development. Doctor Love studied the relative lack of brightness in the different groups and concluded that the truly hereditary cases were most often intelligent, the acquired cases next, and finally the sporadic congenital.

As to the deterrent effect of deafness on the intelligence the question here is one of basal intellectual capability coupled with the retarding effects of a great handicap. The possessor of a highly capable intellect will suffer proportionately much less from any manner of handicap than will the less bright mind. Undoubtedly the developmental value to the child's mind of hearing what is spoken in his environment, and the developmental value of talking, are of very great importance. The deaf child must, then, be mentally considerably retarded, even though possessing a normal mind. The appearance of intellectual retardation is also often enhanced by failure of the parents to teach the principles of self-control and natural adjustment to the child's environment. If neglected in these respects the child may be unmannerly, inconsiderate of others, and uncontrollable. The education which care may afford, in observation and understanding of the child's environment, in self-control and good manners, may do much previous to the school period to help the child over the effects of its great handicap.

DIAGNOSIS OF DEAF MUTISM*

Diagnosis calls for the determination of: First, the hearing; as to (*a*) total deafness; (*b*) hearing for loudly spoken words; (*c*) capability of distinguishing between vowel sounds; (*d*) the presence and degree of a lesser remnant of hearing; second, the condition of the mentality; and third, the physical condition of the ear and its associated throat organs, especially from the standpoint of possible improvement in, or retention of, the remnant of hearing.

The younger the child, the greater is the difficulty of securing its conscious coöperation in carrying out either hearing or speech, tests. With the very young child, patient observation without the child's coöperation will sometimes be necessary. In all cases of mutism in children, however, whatever be the age of the child, the first thought should be to determine whether the child understands and appreciates what is said to him with conversational loudness. The elicitation of this fact ordinarily need not require conscious coöperation of the child. The physiologic significance of the fact in question is this; so far as the education of the auditory word center is concerned, the mute child who hears spoken words is in approximately the same condition as the hearing child who talks. Even at a very early age this center has become impressed with word memories, even though no effort has been made to speak the words so impressed, and the mute child is entirely capable of appreciating and responding to them when they are spoken. An intelligent history is here important. For, an intelligent history must always include an answer to the question, "Does the child understand what is said to him?" Intelligent mothers usually have a clear answer to this question one way or the other. If the child, according to the history, does understand ordinary spoken speech, deafness does not exist, and cannot explain his failure to talk. But if the history displays doubt on this point, or if it indicates indifference to spoken words, or irregularity in his grasping of what is spoken before him, the question of sufficient hardness of hearing to throw a doubt on the clear appreciation of words of ordinary loudness, comes to the front. Further careful history taking may also throw much light on the possible existence of partial deafness

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* The subject of the diagnosis of deaf mutism in some of its various complications will also be found touched upon under the subject of "General Diagnosis of Mutism," and under "Diagnosis," in the section on "Temporary Mutism."

In interpreting the history, as well as in office testing, two sources of error must be eliminated. Lip reading must be rendered impossible by making sure, while the experiments are being made, that either the face is turned away from the eyes of the child, or else the lips covered. And, also, indicative gesture, which the deaf child is especially acute in interpreting, must be entirely avoided. Since the mother is apt not to appreciate either of these important considerations, at least until warned, her history might be quite unreliable. Therefore usually she should be asked to repeat the home experience before the physician, in order that he may judge of their value, or else he must carry out office tests himself.

In office testing let the child be asked in a normally loud voice to do something which can easily be done by his own action, like, "Please shut the door," or "Please see what is happening out the window." If the response be prompt and correct, and especially if similar tests be repeatedly successful, the mutism is certainly not dependent on deafness. If there be no response to such tests, a number of possibilities may explain failures. (1) Marked deafness, or else hardness of hearing; (2) feeblemindedness in a hearing child; (3) although hearing and understanding, bashfulness, intellectual dullness, lack of interest, fright, etc. may deter the child from responding.

Another test having a similar purpose may be employed. With the lips of the physician covered, or else placed directly in relation to the patient's ear, simple clearly spoken elemental sounds like "O," "OO" "a," etc. or simple words are to be pronounced with especial distinctness, and the child asked (by motions as well as by words) to repeat them. In case of continued failure both of these tests should be carried out with a loud voice. The speaking tube will serve both to elicit interest and to help to carry out loud tests.

In case of continued failure, and continuation of doubt in the physician's mind, repetition and continuation of intelligent observation—always attempting to arouse the interest of the child—will ultimately indicate either (*a*) that the child is really deaf, or hard of hearing, or that the mutism must be explained on other grounds.

The testing of the mute child by very loud sounds has but a limited value. (*a*) Since conscious coöperation on the part of the child is usually absent, the interpretation of the child's response, or absence of response, to the sound made, is subject to possible error. (*b*) Physical jar caused by the production of the sound, must be avoided, or discounted. (*c*) Error of interpretation has not uncommonly resulted because of the failure of a hearing child to respond to such tests, because of his childish concentration on some matter of interest, or, possibly, because of his discounting the importance of the sound heard. (*d*) Even though we have become convinced that the child hears loud sounds (and nearly all deaf mutes possess a remnant of hearing) we shall still have to determine the hearing for less loud sounds.

The determination of the exact capability of hearing must eventually be made, as an aid to the determination of instruction measures.

PREVENTION OF DEAF MUTISM

In the direction of prevention of congenital and acquired deafness lies a great hope for decrease of deaf mutism in the future.

Dr. J. Kerr Love, in a masterly series of lectures on this subject recommends for acquired deafness: (1) Ear complications of children to be treated by otologists and to be followed up with prompt effort, persistent enought to bring the best results. (2) Meningitis to be a notifiable disease, and studied until its nature and causes are better understood. (3) Systematic medical inspection and treatment of the ear diseases of children by specialists.

For hereditary deafness: (1) Segregation of the feeble-minded deaf and the prevention of their marriage; (2) discouragement in so far as possible of intermarriage between deaf persons; (3) discouragement, through systematic dissemination of information, of marriage by members, whether deaf or hearing, of any family in which hereditary deafness is actually known to exist.

And as to sporadic congenital deafness: (1) Improvement in the living conditions of the poor; (2) syphilis to be made a reportable disease, and not only the deaf to be treated, but also the parents, and all other children.

TREATMENT OF THE EARS

If the drum-membrane be wholly intact and the deafness profound, the evident location of the impairment in the internal ear makes treatment useless. On the other hand, if the drum-membrane indicates past, or present, disease, a further study of conditions in the ear, the eustachian tube, the nasopharynx and nose should be undertaken, and an effort, dependent on the conditions present, to determine the improvability of the hearing made.

THE RESIDUAL HEARING

A degree of hearing which affords nothing more than a vague perception of very loud sounds can be of little, although perhaps of some slight suggestive value, with reference to the acquirement of speech. But a degree and capability of hearing which permits of the differentiation even slightly of pitch and quality of the loudly spoken voice (with or without mechanical aids) is capable of affording great aid to the development of speech. And if the degree of hearing permits of the actual differentiation of the voiceless (whispered) sounds, and of course of the voiced (loud) sounds, even if accomplished only with marked attention and with the help of artificial aids, and with the assistance of the eye, and even with that of the skilled instructor, the sufferer is removed essentially from the class of the deaf mutes. For the speech is then capable of construction chiefly with the auditory word center as its developmental basis; the child then possesses a subjective differential realization of the sounds he utters; his speech, while modified by his hardness of hearing, possesses a timber and a clearness and accent approaching much more nearly to normal than that of the deaf mute.

Stimulation and Development of the Residual Hearing

In 1893 Urbantschich emphasized the value of stimulating and developing the residual hearing of deaf mutes. His work has been followed by that of Bezold and others. It has consisted in the employment of various instruments for the production of loud sounds of graduated pitch. The patient was expected to differentiate between the sounds of different instruments, and between differences of pitch of the same instrument. These methods did not have the success that was hoped for them, but were not without some degree of success. But they have led to the far more valuable and successful employment of loudly spoken sounds, both with and without artificial hearing aids, in the development of the sense of hearing and of the differentiation of sounds.

The deaf child presumably attends but slightly to the inadquate mental impressions ordinarily made by sounds received in his ears. They are not definite enough to produce the impression required to excite interest and differential understanding. But by making them as clear and definite as possible and by pointing out their language, or other significance, the attention of the child can be attained, and careful action of mental processes necessary for the differentiation between sounds secured; thus, the hearing center is caused to develop its latent function; and the same is especially true of the apperceptive center for spoken sounds (the auditory word center). Even a slight degree of hearing may serve to develop a capability of perceiving at least partially the difference in timber and pitch between loud consonant or vowel sounds, or between the sounds of words as a whole; and a greater degree of hearing may render the complete education of the auditory word center an actual possibility.

Education

In civilized countries a normally minded deaf mute who is totally uneducated is in this day a rare incident. Since the establishment of de l'Epée's School in France in 1755, Heinicke's in Germany about the same time, Braidwood's in Scotland in 1760, and Gallaudet's in Connecticut in 1817, the progressive development of systematic education for the deaf has steadily progressed. All but three, or four, states of this United States are now provided with their own state schools. Public day classes for the deaf are gradually spreading over the larger cities. Many private schools have also been established. One of the advantages of the day classes over the all-deaf institutions is the natural greater mingling of the deaf child with the hearing population. Thus his isolation in a separate social class becomes less conspicuous and less emphasized, his stimulus to talk and speech-read are encouraged, and his later associations with the hearing tend to discourage intermarriage between the deaf. Undoubtedly, however, the all-deaf school possesses advantages also in promoting more constant and systematic and intensive educational effort.

The outlook on life for the deaf mute child depends more than all else on the possibility of intercommunication with his fellows. The ways that are open to him to communicate with others are through: (1) Natural gesturing; (2) the manual alphabet and sign language; (3) actual speech, and (4) writing. He may understand others through: (1) Natural gesturing, (2) the manual alphabet and sign language, (3) lip (or speech, or facial) reading; (4) writing. Each method (excepting natural gesturing) requires the acquirement of an especial system of knowledge. Natural gesturing is too limited in its possibilities to warrant its acceptance as a system of education for any deaf mute whose intellect permits of the acquirement of something better. The manual alphabet and the sign language are not only in general, relatively to speech, limited in their practical possibilities, but they are possible of employment only with the comparatively few individuals who, like himself, have acquired a knowledge of those systems. They have the advantage of being simpler and easier of acquirement than talking and lip reading. But the latter methods permit of communication with all of one's fellows, and in a relatively natural and complete manner. For generations a controversy between the relative advantages of the sign and manual method coupled with writing, and the oral method with lip reading, has gone on. The tendency at present is decidedly towards the "pure" oral method. But a system which combines the two methods also has its merits, its enthusiastic advocates, and wide popularity. A useful remnant of hearing may serve to throw the scale clearly and emphatically towards the pure oral method, which is always the ideal to be striven for.

Too often the young deaf child previous to his attendance at school, because of a lack of understanding on the part of the parents, is left to make his way with much less attention than that given his hearing fellows. Being so seriously handicapped he can make much less progress in the development of mind and knowledge, without assistance, than could his neglected hearing fellow. His handicap calls for especial attention and especial guidance, rather than neglect. The senses of sight and of feeling are most largely to make up for his lack of hearing. He should, therefore, be guided systematically to develop his powers of observation in all possible ways—through pictures, toys, useful bits of work, etc. This training of his powers of observation should also be directed continuously to observing the lips and face and teeth and tongue in talking. Thus he comes to appreciate the use of these organs for talking, and to become interested in them. He should be talked to much as hearing children are talked to, and encouraged to watch the face of the one talking. He may thus acquire the beginnings of lip reading, and his interest be awakened before school age. And he should be encouraged before school age to "talk" himself, that is to babble without coherence much as young hearing children do. He thus becomes acquainted with his speech organs, their sensations and movements. At the same time his sense of touch should be developed along the lines of the best kindergarten instruction. All of this is also making for general mental development, which his hearing fellow is attaining more successfully through hearing others talk and talking himself. Thus at seven or eight years, when school instruction usually begins, the deaf child will be much better fitted for beginning instruction. In some of our cities especial preliminary instruction for the deaf child, during two or more years previous to the regular deaf class age, is being provided. And where this fails, and circumstances permit, especial skilled preliminary tutoring is highly desirable.

Undoubtedly the ideal of educational accomplishment for the deaf mute is that of intercommunication by actual talking and lip reading and by writing. Acquirement of speech by the deaf is accomplished with excellent success by some, with indifferent success by others and scarcely at all by the remainder. With deaf mutes possessed of high mental gifts and afforded the best instruction in sufficient amount, the outcome is all that could be hoped for for these sufferers, both as to talking and lip reading. Such individuals, while always seriously handicapped, may approach fairly closely to a normal, and even cultured life. The less mentally gifted, and the less favored in educational opportunities, have correspondingly less success in talking and lip reading, and experience their handicap in life correspondingly more severely. And still others may make but little progress either in talking or lip reading, and to them the sign and manual method, together with writing and ordinary gesturing, become the only means of social intercourse.

The handicap of the deaf mute is a severe one. He deserves the sympathy and intelligent aid of all the hearing world. If intelligent he is not usually a mendicant, but is industrious and self-respecting and self-supporting. Usually his outlook on life is that of a skilled workman at a trade, or of some useful unskilled occupation with his hands. In some walks of life he may take his place side by side with the hearing competitor, and succeed. In general, however, his handicap demands a certain amount of fellow sympathy, and consideration—a consideration that the world should learn to cheerfully grant.

TEMPORARY MUTISM

(Delayed Development of Speech; Mutism of Hearing; Hearing Mutism; Alalis Idiopathica (Coën); Auditory Mutism)

Physiologic Mutism.—Mutismus physiologica, is the term applied to those cases in which the speech develops normally, but begins late in the normal period. The speech in such cases usually goes on to complete normal development. The lateness of inception of the speech processes, if inception occurs within the third year, is of little significance, excepting for the doubt engendered by the delay that the child may in some manner not be normal. If the delay continues into the fourth year, this doubt becomes fully justified. Normally constituted children, who have suffered prolonged illness, are commonly subject to such delay in speech beginnings. Boys are more often delayed than girls. The general and particular normality of the child is the only guide as to the normality of delay in speech development, and if doubt exists on this point the matter should be medically investigated.

The true temporarily mute child is, as a matter of course, not at the time entirely normal, although the approach to normality, both mentally and physically, may be close. The condition represents a pathologic status of delay in the action of one of the fundamentally important human functions. In proportion as this abnormal delay continues into the normally speaking years, the mental growth of the child is impeded by its relative mental inactivity due to the absence of speech. In other words delay means mental retardation, even aside from the causes which may have produced it. With these children, if left to themselves, some manner of speech activity is seldom delayed beyond the eighth or ninth year, although Gutzmann records a case of spontaneous inception of speech at 15 years. This fact, that the speech may usually be expected eventually to spontaneously make its beginning, does not, however, render the subject unimportant. The significance of delay in the beginnings of speech development arise from the diagnostic doubt as to the seriousness of the cause of the retardation, and from the doubt of the normality of progress in speech development and of its eventual normality of outcome, once it has begun. For, the child whose speech is abnormally late in appearing is very likely never to spontaneously develop normal speech, and may not succeed in such an outcome even with expert assistance. Moreover, marked lateness in beginning speech, especially if the speech once begun be developed slowly, constitutes a serious hindrance to the entire process of mental development. Such a child, for example, even though he be essentially normal in mentality, may at eight be compelled to occupy itself with much of the developmental activities of three, or four, or five. He thus is always behind with respect to the grooves of life laid out for normal children, and, therefore, is ever in need of especial and expensive attention and instruction.

The temporarily mute child may, as to mentality, be moderately, or slightly feeble-minded, or he may be merely mentally backward, or he may be essentially normal; but, in any case, so long as the mutism exists, the mental processes of such a child are not functioning with entire normality. The child hears and understands spoken speech; therefore the receptive word center is in the process of education and development, although handicapped by the speechlessness. The child may seem stupid, or ordinarily bright. He may speak crudely a few simple words, or none at all. He may make crude inarticulate sounds which with a gesture may serve many purposes of self expression. He may have begun to talk and then have suddenly ceased. He usually employs gestures for his language needs and may employ them intelligently. Indeed one cause for his failure to essay to talk may be his skill in the use of gestures, and especially the misguided acquiescence of his parents in that skill. The more useful expenditure of childish energy in the acquirement of speech may thus be wasted on the acquirement of gesturing. He may seem to try to talk but produce only unintelligent jabber, or he may seem not to have the remotest thought of talking.

A certain manner of bringing up of these children not only complicates the difficulties of diagnosis, but also complicates the treatment, and creates a false relationship between parent and child of serious consequence to both. I refer to the misguided indulgence of the unusual child, i.e., the failure to educate the child to a sense of obedience. Such a child being dominated by childish wilfulness, and being out of normal relationship with his environment, may have many of the characteristics of an unguided animal, and become a serious problem from the standpoint of self-control alone.

Etiology.—The section above on the "General Etiology of Mutism" (p. 746) will serve to point out the backgrounds of the etiology of temporary mutism. And the remark heretofore made may be repeated with a modified application, namely, that the incentive to talking is so strongly rooted and so instinctive in the child that its marked delay in beginning must usually have also a relatively profound cause.

And this cause is nearly always at its root psychologic. Peripheral physical handicaps are not uncommonly present, to be sure, but whether they operate without an accompanying causal psychologic perversion, is at least open to doubt. For speech usually does make its beginning and does progress in normally minded children in spite of serious and varied peripheral handicaps. For example: Gutzmann thinks that marked overgrowth of adenoids may be sufficient to inhibit the incentive to speech. This does not seem unreasonable, especially if one speaks of short periods of inhibition; and yet Liebmann finds no instance in these children in which the adenoid operation has served to stimulate spontaneous speech. But if adenoids served to induce a relatively profound mental retardation, as in "aprosexia," their influence in the retardation of the beginnings of speech would seem probable, for the reason would here be also psychologic. And in the instances mentioned also by Gutzmann, in which adenoids are associated with marked hardness of hearing, the physical handicaps might be so severe as to fully account for the prolonged retardation of speech. But even here the cause might be considered mental rather than physical, since the receptive word center could be but imperfectly educated and stimulated. Relative immobility of the tip of the tongue (tongue tie) is entirely insufficient of itself to be causal. Gutzmann mentions cleft-palate as a possibly sufficient cause, but only the rarely exceptional cleft-palate child fails to develop some manner of speech. Occasionally a child who is severely paralyzed in the limbs and even slightly in the tongue, comes in this class, but in the only case of the kind I have observed a child who began to talk at six, the complete normality of the mind was in question. Prolonged illness also acts in the retardation of the onset of speech through impairment of the mental as well as the peripheral physiology of the child.

Heredity as to mentality applies in certain instances of feeble-mindedness, and possibly even of certain of the mentally dull. Capability of attention and of concentration are commonly impaired. Lack of skill and aptitude in muscular movements in general has been noted (especially by Gutzmann). Liebmann notes lack of capability for observation, and also impairment in the action of different sense activities. The influence of alcohol on the child is emphasized by Coën.

Birth traumatisms in the head region are quite frequently noted in the history of such cases, and in certain of this class injury of the motorwood center is probable. The mental effects of marked bashfulness has been mentioned by Nadoleczny as a possible cause. In connection should also be mentioned the effect of harsh and tactless environmental conditions on the sensitive child, especially if the mentality be backward, or the child extraordinarily sensitive.

Summing up our knowledge and speculations as to the causes for delayed onset of speech (temporary mutism) one finds clearly physical explanation in the feeble-minded cases, the paralyzed cases with mental impairment, and the child in ill health. He finds at any rate partial physical explanation in the hard of hearing, the paretic, the tongue tied, etc. But in a portion of these, and in another large group, the explanation is to be sought in a variety of psychologic causes, often exceedingly obscure.

Diagnosis.*—The temporarily mute child must possess hearing sufficient for the understanding of speech, peripheral speech organs capable of performing the physical acts of speech, general mentality sufficient for speech requirements, and fairly competent speech centers. Deaf mutism must be excluded. To differentiate between profound deafness and marked hardness of hearing in the mute child may require ingenuity and much observation. The word of the parents on this matter is important. Determination of the exact degree of hardness of hearing will be an advantage when instituting treatment. Marked feeble-mindedness must be excluded as offering no hope of more than the simplest and crudest speech. In view of the fact that, even in the cases that eventually prove their essential mental normality, nevertheless the mind of the mute child shows peculiarities and retardation. In view of the speechlessness and general lack of coöperation of the child, the determination of the exact condition of the mentality, whether moderately feeble-minded, or backward, or normal, is sometimes extremely difficult, and may require long observation. The interest in and reaction to pictures is to be noted, also the manner of reaction of the child to different kinds of play. If possible the child is to make attempts at imitation of spoken sounds or words, constituting the most definite and practical test of the capability for talking, and a valuable test of attention and concentration. To accomplish this important test may require days of play and tactful handling, in order to gain the confidence and coöperation of the child. If one succeeds in obtaining good imitative efforts the capability for the acquirement of some manner of speech may be considered established. Borderline mental cases may not be certainly diagnosed as to the mentality until after treatment has been instituted for a considerable time. Injury to the motor word center would be likely to be accompanied by paralysis, but by no means all paretics are aphasic. Injury to the sensory wood center is excluded by the readiness with which the child understands spoken speech.

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* The reader is also referred to the section on "The General Diagnosis of Mutism."

Prognosis.—The mute child who readily understands speech, who is in good general health and whose mentality is normal, or close to normal, usually is capable of speech. Whether the speech shall become completely normal depends on the age at which speech is begun, and, in a large percentage of cases, on the treatment which is instituted. The mute child with hardness of hearing requires much careful and skillful technical instruction, if normal speech is to be arrived at. The slightly feeble-minded mute child is apt to attain to a more or less imperfect speech, dependent on the peculiar mental qualities of the particular child, and the treatment instituted. The speech attainment in cases of other complications is dependent on the individual abnormality.

Treatment.—The first consideration is the attainment of a normally acting general physiology. This is of especial importance if sickness or malnutrition are producing physical and mental depression. For one may commonly expect under such conditions neither a spontaneous effort towards speech, or success even with helpful aid. Of course, however, if the general health necessarily remains impaired for some months, an undelayed effort should be made toward stimulating an effort at speech production.

The degree of suspected or determined mental impairment must have a bearing on one's attitude towards the child's speechlessness; for, while marked mental impairment might deter one from hoping for the attainment of speech, a lesser degree of impairment would call for the testing out of the situation through attempt of aiding the child; while slight feeble-mindness might call for painstaking efforts to arrive at the best possible speech.

The condition of the peripheral speech organs should be given attention. The most important disorders requiring correction are *marked* overgrowth of adenoid tissue, *marked* impairment in the possibilities of movement of the tongue by reason of very short frenum and, very rarely, cleft-palate. But let it not be forgotten that the child is not usually deterred from *attempting* speech by purely local peripheral abnormalities, and that such conditions are usually merely accessory to more subtle psychologic or physiologic explanations.

If, with or without correction of peripheral abnormalities, the child has failed to show signs of attempting spontaneous speech, efforts should be begun towards arousing spontaneous effort in this direction. The basal purpose is to endeavor to develop from within the child's mind a *desire* to attempt to use its speech organs in talking. The child is usually stolidly indifferent and unappreciative of speech. Force and insistence do harm. The round about way, if it be tactful, is the most successful. The childish nature and his pleasures are to be appreciated and taken advantage of. Underlying the efforts of the instructor, should be a knowledge of physicologic phonetics. Gradually gaining his friendship and confidence he is to be tactfully led to play with his speech organs, blowing, puckering the lips, sounding whistles, blowing out simple sounds, like ō ōō, etc. He is to be taught to observe the lips of the instructor as talking goes on. Picture cards and blocks with letters or words printed on them are valuable aids. All the while he is to be educated tactfully to obedience, while still retaining his confidence and friendship. More direct imitation of words is to follow. If the child be sufficiently capable mentally the inculcation of a knowledge of writing may vary and support the work with the speech itself.

As he becomes more tactable the substitution of single words to take the place of gesturing is to be quietly insisted on. Thus gradually a beginning vocabulary is formed. Beyond this some children proceed unaided, but most of them require much help over the difficult sounds and words. They then come into the class of sufferers from articulative defects treated in another chapter.

When one considers that in the natural course of events a certain child should have begun to talk spontaneously, and when it has become clear that the environmental attitude towards that child is not conducive to its psychologic normality, a separation of the child from its home environment, or at any rate a certain manner of reform in that environment, may be necessary in order to bring about success in developing speech.

DISORDERS OF ARTICULATION

DYSLALIA; STAMMELN *

The outward form and character of the developmental disorders of articulative speech readily suggest themselves by the crudities of speech of the developing normal child. They are analyzable into two great divisions, one concerned with the production of the individual word (aphasic) and the other with the production of the sentence (agrammatic). Between the beginnings of speech in the young normal child and the attainment of complete speech, one views a gradual progression, representing the advancing success, step by step, as the mind develops and the skill for speech production increases. Each period of progress presents its peculiar characteristics. Not until about the third year does the child graduate little by little from the one word manner of expression, to sentence building. The articulative disorders proper present themselves as more or less exact reproductions of some stage of the physiologic dyslalling. The physiologic dyslalling thus remains to a large degree unprogressive, and, therefore, pathologic. The agrammatic disorders of speech development, while not truly articulative, are always associated with much dyslalling and are usually dependent on the same causes, and, therefore, are best considered in close relation to the articulative disorders proper.

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* The long employment in the German language of the word *Stottern* to signify the speech disorder designated in the English language by *stammering* and by *stuttering*, and the absence of any popular English word to correspond in meaning with the German *Stammeln* has led to and is still causing confusion among English writers on speech disorders. What is needed is an agreement among students as to a newly created English term to serve the very useful purpose of the German word *Stammeln*. The employment by certain American students of the English

The progression of the child to articulative perfection concerns itself with successive, oft repeated, efforts to produce more and more accurately the combined laryngeal and oral positions upon which each elemental speech sound depends, and upon increasing the facility and accuracy with which these elemental positions are combined to produce words.

The efforts involved concern the appreciation and apprehension of the sounds and words heard, the mental effort required for directing their reproduction, and the complicated muscular action of the peripheral organs of speech. In the course of the years required for the complete development of these complex processes two great speech and memory centers become permanently educated, that for apprehending spoken words, and that for reproducing them. At any particular stage in the development of the speech these memory centers are correspondingly made up of crude and partial images. If the processes of speech development cease to progress and a particular sound position continues to be permanently reproduced in an imperfect manner, the memory centers for that sound have become wrongly educated. The same is true for combinations of sounds. If particular elemental positions each representing an individual sound had never been produced at the time that progress in speech development ceased, no memory for that sound comes into existence, and the speech must be permanently constructed without it, or at any rate, until, through training, it has been supplied. Either the undeveloped sound is

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word stammering, which for untold generations has stood for one definite meaning (also designated in large part by the word *stuttering*), in the meaning of the German *Stammeln*, cannot be too strongly condemned. Such an artificial alteration of the idiopathic popular meaning of any word is impossible. The result can only be the creation of perpetual confusion as to the meaning of the term whenever it is employed. The whole difficulty can be fully and satisfactorily solved by the agreement on a certain new word to cover the meaning of the German word *Stammeln*. For this purpose the writer would suggest the word *dyslalling* (Greek, *ill*, and *talk*). The verb would be dyslal, dyslalling, dyslalled, and the adjective dyslallic.

omitted from words, or, more frequently, another sound is substituted for it.

This permanent maleducation is proven for the kinesthetic (motor) word center by the continuance of the wrongness of production of the word by the peripheral organs of speech. A doubt might exist as to the corresponding maleducation of the sensory word center. For it might be argued that the malproduction of a word or sound were due to conditions having no reference to the receiving sensory word center, which, therefore, might continue to possess correct sound and word images; and, at the beginning, particularly in cases where peripheral organic malformations were largely responsible for the defect of articulative position, this might be true, and probably often is true. But as time goes on and the rightly apprehended word is "corrected" by receiving back over and over the sound wrongly produced from the peripheral organs of speech, even the sensory word center becomes itself wrongly educated. That this is true is evidenced by the complete acquiescence of the individual in the wrongly produced word, and even in his failure to realize his own lack of correctness. In most cases, however, it is certain that the malaction of the peripheral organs of speech is immediately dependent on the imperfect action of the sensory word center, the imperfection in the motor center being thus quite directly dependent on a primary impairment of the action of the receiving center.

In many cases, if not in all, the failure to progress in speech development is dependent, at least in large part, on alack of attention and concentration on the problems of exact word imitation. One finds not alone inaccurate sound production, absence of individual sound production, and substitution of one sound for another, but he finds, with complete regularity, a failure in the midst of words and in the ending parts of words much greater than in their beginnings. This may be due to more than one cause, but it is probably due most of all to the greater intellectual effort required to clarify the image of words as one proceeds beyond their beginnings, and to carry out accurately a more prolonged peripheral effort in producing the word.

To what degree and how universally the articulative developmental disorders are to be considered a measure of general intellectual impairment has been quite frequently discussed. It may be said without question that the degree of dyslalling is in a large percentage of cases a fair measure of intellectual impairment, but it is just as truthfully said that also in a large percentage of cases individual conditions of temporary ill health, of environmental care, of peripheral disturbance, and of bad examples of speech, are largely determining factors. Children who have grown even to maturity with a practically nonunderstandable speech have had the disorder corrected and have taken positions in life requiring high intellectual capacity.

Sentence building implies the grasping of a thought to be expressed and the combining of words for its expression. These are both intellectual processes, and sentence building is generally speaking a genuine measure of intellectual capability. It is therefore in the feeble-minded that this defect becomes especially pronounced.

Etiology.—The causes which produce temporary mutism, are the causes also of dyslalling. In qualification of that statement, however, it must be said that causes, both functional and organic, of so slight a character as to bear no evident relationship to causes which produce mutism also may produce articulative disturbance. The causes are commonly apparently functional; they may be organic, but even when organic, a functional element often seems to play a part. The most evident cause of articulative maldevelopment as well as of failure in sentence building is feeble-mindedness. Also mental backwardness is found in a large number. Nearly all the cases of temporary mutism enter, when they begin to talk, into the category of dyslallics and agrammatics. In some cases it seems quite clear that the sensory word center is obtunded in much the same manner and degree that the musical "center" is functionally impaired in persons who possess very little "ear" for musical pitch. Theoretically the motor word center might be similarly obtunded.

On the peripheral side we must especially note hardness of hearing, cleft-palate and other palatal disturbances, deformities of the teeth, short lingual frenum, and paresis of the tongue and lips. Certain of these organic causes will require separate presentation.

Manifestations.—The most common articulative disorder, and, indeed, the most common defect of speech, is *lisping.* In the production of the s sound the tip of the tongue rests lightly behind the lower teeth, the forward half of the tongue arches upwards, and while shutting off all air at the sides, forms a central anteroposterior groove on the upper surface, which directs the outgoing air against the upper teeth. Another manner of production of this sound, much less common, but producing essentially the same result, consists of the placement of the tip of the tongue directly against the lower border of the upper teeth, this forming a groove for direction of the outgoing air against the upper teeth. The most common manner of lisping is that in which the tip of the tongue projects more or less completely between the teeth, the air escaping between the upper surface of the tongue and the lower border of the upper teeth. This is the normal position for the th sounds, and this manner of lisping (parasigmatismus; sigmatismus simplex) consists really in the substitution of the th sound for the s. The causes may be entirely functional, or they may rest upon deformities in the approximation of the upper and lower jaws anteriorly.

Another manner of lisping consists in the escape of the expelled air through a lapse in the approximation of the sides of the tongue to the upper teeth (sigmatismus lateralis). The result is a rough, harsh, crude s sound that constitutes a serious speech imperfection. Here the cause may rest on misplaced, or markedly irregular, teeth, or absence of certain teeth in the upper jaw, or a markedly misplaced anterior approximation of the jaws. Another and still more serious form of lisping, more serious because the imperfection of sound is louder and more disagreeable than that produced by either of the other two, is the nasal lisping. This results from functional inaction of the soft palate for this sound. The front of the tongue usually assumes the normal s position. But the base of the tongue raises and is approximated to the lowered soft palate. The air for the s sound passes through the narrow space between the base of the tongue and the lowered soft palate, sets the latter into vibration, which vibration possesses a strong nasal resonance. The sound suggests a pig's snort. Relatively this is a rare defect. Since the z sound requires the same oral position as the s, it also partakes equally in any defect in the s sound. The sh and zh, which are formed similarly to the s and z, may have similar defects in association with those of the s and z.

One or two defective sounds, as, for example, that of a lisping s and z, may constitute the entire articulative defect. But even one defective sound, since it occurs and re-occurs through many words, may vitiate the entire speech. From 1 to 15, or even more, elemental sounds (out of 45) may fail entirely to be produced. Malformation of as many other sounds may also be present. In addition a certain number of substitutions of one sound for another may occur; and finally, the entire speech may be vitiated by general inexactness, this applying especially to the central and end parts of words. The sounds most commonly involved, either by omission or malposition, are s, z, l, r, k, g, ch, j, but no sound is incapable of malproduction. The speech may be as understandable as if normal, although presenting defective peculiarities, or it may be so distorted as to be practically non-understandable; and intermediate states in all degrees occur.

The dyslalling of the feeble-minded presents various degrees of imperfection. The especial peculiarities are dependent naturally upon general defects of intelligence. The entire picture is likely to be one of marked vagueness of purpose. No sound may be accurately produced; unproduced sounds are likely to be numerous, substitutions are common, the ends of words are elided or produced with extreme carelessness. And it is in the feeble-minded that sentence building is most likely to be imperfect (agrammatismus.)

Shortness of the frenum of the tongue is never of itself sufficient to prevent the development of speech, although it may theoretically in extreme cases, especially if natural aptitude for speech production be lacking, discourage the child from efforts at speech production. Its effects in producing articulative disturbances are not highly important in the normal child. Such a condition may in a child be susceptible to functional disturbances of articulation, discourage the production of the t, d and n, or cause them to be produced carelessly. When the tongue is clearly hampered in movement anteriorly it should be given more movement by severing the frenum, but one should not preseume that such an operation will of itself without training produce great improvement in the speech.

The dyslalling of the hard of hearing presents on close observation rather definite peculiarities. This cause operates only when the degree of deafness is sufficient to produce impaired sound images of spoken words, and consequently a primarily wrongly educated sensory word center. The defects concern naturally much more the voiceless (breath, or whispered) sounds than the voiced (loud) sounds, for the voiceless sounds may never have been clearly heard. The ends of words are likely to be especially obscure and may be omitted altogether.

Prognosis.—For the normal child the prognosis is excellent; for the feebleminded child it is dependent on the degree of feeblemindedness, but is always bad; for the backward child much can be done, but the degree of attention and concentration that can be secured determines the outcome. Lisping can usually be corrected even without correction of any present deformities, but more readily after their correction and sometimes not without their correction. Defections due to hardness of hearing are correctable through the employment of artificial aids to the hearing, if necessary, to bring to the child a perfect apprehension of the sound and word to be imitated.

In the child who may chance to acquire speech with difficulty the employment of baby talk in the presence of the child is a serious mistake. Indeed it is never a safe practice with any child. Prophylaxis calls in general for the finest examples of speech, spoken slowly and distinctly. If the child be hard of hearing adequate loudness of speech is also necessary. Compulsory methods as a means of prophylaxis do not usually work well, but in the child too young for more elaborate training slow and distinct speaking before the child will do much. A tactful guidance with respect to especial difficulties carried out for short intervals, many times repeated, is in many cases successful as a measure of prevention.

Treatment.—It is always to be understood that physical handicaps, whether affecting the child mentally or peripherally, should so far as possible be removed. This remark applies to conditions of ill health in general, whatever their character, for such conditions necessarily affect the normality of cerebral physiology. Adenoids, if large, may enter into this class. Adenoids may also be an important factor in the causation of the middle-ear deafness which lies at the root of certain instances of dyslalling. And more than this adenoids may have much the effect of a foreign body in hampering the movements of the palatal muscles and of the salpingo pharyngeus, which is inserted partially in the thyroid cartilage and hyoid bone. Moreover, adenoids, acting also as a foreign body, serve to disturb the normal muscular and mucous membranous sensations of the pharynx, upon which sensation the delicacy of peripheral speech action largely depends. Removal of adenoids, then, is a matter for consideration in any individual case of dyslalling in which they exist. Only when the faucial tonsils are believed to be a factor in producing impairment of the general health, or when they are greatly hypertrophied, need their removal in dyslallic children be given consideration.

Deformities of the jaw and teeth, which render the production of certain sounds, especially the s, z, sh, and zh, more hard to produce, are likewise in the individual case subjects for especial consideration. With a bright child, however, only rarely do such deformities actually prevent the production of any sound.

If the tongue can be protruded well out of the mouth, and especially if the tip can be at all curled upwards over the upper lip, and even if the latter of these two acts be accomplished only imperfectly, the movement of the front of the tongue may be considered sufficient for speech purposes.

As early as is possible in the individual case, the training should become definite. Each unformed elemental position is with the aid of the eye, by given example, and with the use of a hand mirror, and also by clear sound examples, to be tactfully and patiently taught. Individual sounds which are imperfectly produced are to be corrected in a similar manner. All possible combinations of these newly formed sound positions, and of corrected articulative positions, with other sounds, are, as they are realized and quite correctly produced, to be made and practiced many times over. Remembering that the speech habits both central and peripheral have long been forming the necessity of care in practicing, especially the carefulness engendered by the constant imitation of correct sound examples, and by the use of the hand mirror to observe the articulative movements, will be realized. The replacement of these wrong habits by correct ones is a labor requiring much time and patience. A psychologic danger is to be ever in mind and constantly guarded against. This is the inculcation of a spirit of resistance or even rebellion, on the part of the child. The childish nature is to be tactfully considered. The spirit of playfulness, and a certain amount of circumlocution, for example with blocks and cards and pictures, while having the main purpose always in view, will succeed best with the young child. With older children, or adults, especially in cases in which sound confusion and carelessness pervades much of the speech, it will be best to teach by eye and ear all the elemental sounds of the language, and also the building of any word on the basis of its physiologic elements. In this manner corrections within the word and ending the word may be accurately accomplished. The analyzation of the written word into its sound elements should be a constant accompaniment of this plan of training.

With backward children indefinite patience is required, and sometimes only partial success is to be expected. With the feebleminded at the best only improvement is usually possible. And, indeed, whether any effort, even for improvement, should be undertaken, must be determined in accordance with the mentality of the child and its social circumstances. The amount of time necessary to be consumed in order to accomplish even a slight degree of improvement, and that, too, in a child who is necessarily to be largely isolated and protected for life, is such as to often cause serious question as to its desirability or warrant. It will usually be found impossible for the feebleminded child to acquire the sound positions not acquired spontaneously, and often impossible to make great improvement in those that are incorrectly formed. So the incapability of grasping the problem, the lack in capability of attention and concentration, and the inadequacy of the memory centers will all serve to render such accomplishments difficult if not impossible. Usually the best that can be hoped for will be a certain general improvement in accuracy of enunciation, and specific correction only in the more marked and easily improvable imperfections.

In the backward children and in so far as seems possible and desirable, also in the more nearly normal feebleminded, the problem of encouraging the building of sentences will also sometimes be present. Patient repetition of simple sentences, such as those to be found in school readers or sentences especially constructed for the purpose with the aid and encouragement of pictures and of play things, is to be employed, as well as the tactful, repeated correction of errors in talking.

In cases in which paresis in the peripheral organs of speech is present, practice in movement of impaired organs, aside from speech production, should be given especial weight. Otherwise the methods as outlined above, with due consideration of the peripheral impossibilities, are to be employed; the degree of success will be dependent on the seriousness of the peripheral handicap, and on the degree of mental impairment, so often an accompaniment of peripheral paresis. If spasticity be present in the peripheral organs of speech, the problem of correction offers the additional difficulty, that attempt at definite movement throws the involved muscles into spasm. But if such children be mentally competent patience and gentleness will succeed in bringing about a certain but limited articulative improvement.

STAMMERING*

(Stuttering; Stottern; Balbuties)

Definition.—Stammering is, on the one hand, a spasmodic perversion of the peripheral movements of speech occurring only during talking, and, on the other, a manner of psycho-emotional disturbance intimately associated with the peripheral disturbance; beginning typically in early childhood and tending to gradually increase in intensity usually until some time in early adult life, and to gradually decrease in intensity with the passing of the adult years; and manifesting typical and marked variation at different times in both frequency and intensity of manifestation. These factors of (*a*) peripheral disturbance, (*b*) emotional disturbance, (*c*) gradual onset and gradual disappearance, and (*d*) marked variability of manifestation, must be included in the usual and typical picture of the disorder.

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* The term *stammering* has by usage come to be applied in a broader sense than *stuttering*, i.e., to be of the two terms the more comprehensive. Descriptively in a more restricted use of the two words, *stuttering* has come to refer to the lighter manifestations of the disorder, as evidenced by repeating the initial sound of the word, and *stammering* to the more severe spasmodic manifestations. In the German language the words *stottern* and *stammeln* have long been in popular use, the first referring to stammering, and the second to non-spasmodic articulative disorders producing wrongness of pronunciation, and having no resemblance to stammering. The effort on the part of certain authors writing in the English language to rule out the word *stammering* and employ *stuttering* instead, is an

Symptoms.—Objectively stammering consists of a conscious struggle on the part of the stammerer to produce a smooth utterance, against innate tendencies to resistance and holding on the part of certain of the speech muscles. Consequently the production of speech seems to be uncertain, and instead of flowing smoothly, is broken up into separate successive struggling efforts. And paralleling this peripheral struggle is also an inner psycho-emotional perturbation, of variable degree, but often of the severest character. On the one side, then, is mental disturbance and on the other muscular tensions and spasms and misdirections.

Peripheral Phenomena.—The outward manifestations of the disorder are shown first, to the ear, by perversions of the sounds of the words of speech, and, second, to the eye, by visible manifestations of holdings and distortions in the facial area. What happens in the peripheral area is this: When the stammerer starts to talk certain muscle groups overact, becoming very tense or even entering into firm clonic or tonic spasm—like holdings. This localized tenseness throws the entire sub-consciously acting speech machine out of the possibility of normal operation, and, if the spasm be severe, temporarily out of action altogether. Tenseness of a more general character is apt to pervade the entire body but tends to be more marked about the throat and chest. If the "spasm" be severe relief from the particular manifestation may come only with the demand by nature for breathing. The location of the spasms is most likely to be in the larynx—base of tongue region, but often also in the mouth structures, as well as the chest.

Accessory spasmodic movements, or holdings, or "spasms," involve also, but only in the occasional stammerer, other parts of the face and body, including even the arms and legs. These may well be called "superimposed," or "accessory," movements—the "mitbewegungen" of the German writers. In view of their apparent pseudo-voluntary, or semi-voluntary character, and their occurrence

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attempt primarily suggested by German writers to employ an English term meaning stammering that resembles in sound and written form the German popular word *stottern*. But the effort seems ill advised. The word *stammering* is too deeply rooted in English usage to be discarded, and that stammering could ever acquire the meaning of the German *stammeln* is unthinkable. Popular language is not thus artificially altered, even if it were desirable to attempt it. The wiser plan is to adhere to the long rooted usage of the words *stammering* and *stuttering* and to coin for popular use a word corresponding in meaning to the German stammeln, always remembering that the English *stammering* and the German *stammeln* have entirely different meanings. As a popular term applied to articulative disorders, and corresponding in meaning to the German *stammeln*, the author would suggest *dyslalling* (Greek, *ill* and *to talk*).

beyond the field of action of the speech apparatus itself, the phrase "superimposed movements" would seem to be especially appropriate.

The effect on the speech movements of these muscular overactions is to cause (1) a spasmodic repetition of the first elemental position of the word, or more rarely, of the first syllable of the word; or (2) the quite firm holding of the first elemental position for a considerable period, with or without much audible result. Sometimes the discouraged sufferer, knowing the struggle that attempt to talk will inevitably produce, merely stands, like one stupid, or dumb, without making any effort. Having succeeded in overcoming the spasmodic holdings at the beginning of a particular word, the speaker runs on, possibly hesitating frequently, or only occasionally. The hesitations are more likely to occur on the first sounds of the first word of a sentence. They may, however, not occur for an indefinite number of sentences, or may be repeated on nearly every word.

The susceptibility to muscular tension and the degree of tension and severity of the spasmodic holdings varies much with different stammerers, and also at different times with the same stammerer, or may be temporarily almost in abeyance. A confirmed stammerer may, then, stammer with frequent and intense holdings, and at other times, under different circumstances, may stammer less frequently and less intensely, and at still other times may not stammer at all.

Mental Phenomena.—The most evident fact about stammering is its outward spasmodic interference with the speech, but the really most significant fact is the inward disturbance of the emotions. The *misunderstanding* of stammering is the understanding which reaches no farther than the outward clinical manifestations. It is this superficial *misunderstanding* which alone can account for the relative indifference of physicians and parents to the oncoming of so important a disorder. A complete clinical picture is obtainable only as the emotional picture is made an intrinsic part of the outward spasmodic struggle. And such a complete picture is obtainable only as the inward emotion is shown in its culmination in mental distress and suffering, and only as the combination of spasmodic disturbance of speech, the emotional disturbance and the mental pain, are viewed as crippling the individual socially and economically through many, or all, of the years of life. Strangely hampered in the most human of all desires, the desire for adequate self-expression, the stammerer becomes a prey to his own torturing emotions; misunderstood, ridiculed and misjudged, seldom able to do himself justice before others, he stumbles and falls and recovers and falls again, as he goes on his road of inward distress through life.

The temperamental emotionalism behind a large proportion of stammerers is not without importance in itself alone. When existing without the speech defect such emotionalism is of great importance, but of no greater importance than for the thousands of non-stammerers of like constitution. But unusual temperamental emotionalism when compelled to bear the incubus of this wracking speech disturbance produces a serious situation. The degree of this seriousness is dependent on the degrees of emotional susceptibility and the seriousness of the speech disturbance.

Emotion in Childhood.—Children are characteristically both highly emotional and highly sensitive. Emotional disturbance is, I think, always present even in the youngest stammerer. But the emotional suffering of young children from stammering is difficult to estimate. Especially if the beginnings are slight the child experiences little or no distress for some time. But as the years of the young stammerer go on, and the frequency and intensity of the hesitations increase and he comes to realize the smiles and jokes of those about him, the characteristic mental distress and strain gradually become established. Perhaps a false and morbid conviction of his own peculiar difference from other children comes in to pervade and pester his mind. He tends to hold himself aloof from others, especially in the larger social activities. He attempts to conceal both his emotion and his suffering, and so silently and painfully stumbles and struggles along. In his higher school education the painful struggle continues. And when the time comes for his entrance into the world he faces not only a life of emotional pain and of struggle, but also the possibilities of a life of relative economic failure.

The Beginnings of Stammering.—In probably more than 90 per cent. of the cases no obvious explanation of the onset is perceivable. It lies concealed in the psychologic and physical life of a developing child of three to six or eight years. The first efforts at speech have been passed over successfully. The child is usually in the very midst of the uncertainties of speech development. The disorder is developmental both from the viewpoint of the psychology of the child, and from the viewpoint of the crude and uncertain stage of the mental and peripheral physiology of speech. The disorder arises when the mind and emotions of the child are strongly impressionable by causes so slight as not to affect one of more mature years, and when the education of the mental and physical mechanism of speech is still in an uncertain, wavering, and progressive stage. It is then that stammering obtrudes itself into the developmental insecurity and gradually establishes itself as a distortive perversion of the speech function. The beginnings of the stammering may be so slight as to fit readily into the appealing childish life and conduct, and thus for weeks or months produce no apprehension. The child may repeat the first sound, or he may hold slightly or firmly on the first sound, seeming to struggle to make progress. Or he may violently and evidently struggle with his speech, with much violent and obvious holding. In a general way the extreme manifestations are more likely to be temporary, but any manifestation may be the forerunner to chronicity. Periods of temporary quietude, or even complete abeyance, may occur, only to bridge over the advent of a further attack. The frequently unobtrusive onset is worth especial emphasis. For, as a matter of professional and lay education, both physician and public should become impressed with the importance of prevention, or, at any rate, of treatment before the disorder has become firmly established.

It is often stated that these manifestations occur only in children who are especially excitable, that is, "nervous." Since, however, young children are by reason of incomplete development always "nervous" one has to be careful with respect to such a conclusion. Certain it is that stammering develops commonly in children who appear to be in every way mentally and physically normal. Circumstances which render the child more emotional and self-conscious tend to produce an increase in the abnormal manifestations. When he first enters school an increased intensity and increased frequency of the stammering is usually noticed. So characteristic is this that the stammering may even not have been clearly realized until school attendance was begun. And in a goodly proportion of the cases the very onset itself is directly referable to the emotional excitements of the new and wonderful and trying experiences of the first weeks of school life.

Onset of stammering through association with another stammerer although relatively rare, is of course a phenomenon of importance. The comparative unsusceptibility of the child to this manner of "psychical contagion" is shown conclusively by the great frequency with which stammering develops in but one member of a family of from two to several children. And yet, since the mind of the young child is normally concentrated on the acquirement of speech through imitation, certainly imitation of stammering seems natural; and, also, since every child is emotionally impressionable, the acquirement, along with imitation, of much the same psychical state with reference to speech as the original stammerer, seems far from impossible. However, a certain manner of suceptibility may be required for the phenomenon of imitation to go on to completion, which would account for the relative rarity of imitation. The dangers of imitation should in any case always be appreciated.

In a third manner of onset in children the stammering follows suddenly from nervous excitement, or shock. Fright is the most usual form of emotional shock resulting in stammering. For example: A boy of nine was walking in the dark along a raised sidewalk. Accidentally he stepped off the side of the walk and fell a distance of 2 ft. Although physically uninjured he stammered from that time on. Excepting as one realizes the acute emotional susceptibility of children, the degree of shock is often out of all proportion to the real seriousness of the accident.

Nervous shock continues to be the exciting cause of stammering well into the adult years. During the recent World War, stammering has developed among the soldiers in action in considerable numbers. Cases developing after puberty from any cause are rare, and especially rare from causes other than shock. I have witnessed one clearly defined case, in which the stammering seemed to be built on a constitutional tendency to muscular hypertension with irregular twitching movements, coupled with strain in making a living.

The disorder occurs only rarely among the feeble minded, and the mentality of stammerers is on the average rather high. A goodly number of famed and brilliant men have been stammerers. Boys are several times more susceptible than girls.

Etiology.—The cause of stammering is still a subject of controversy. The discussion involves chiefly the origin and nature of the mental disturbance, and of the peripheral disturbance, and especially the question as to whether the central, or the peripheral, disorder is primary. Kussmaul taught and his pupil, Dr. H. Gutzmann, believes, that "stammering is a coördination neurosis, resting on a congenitally excitable weakness of the articulative apparatus." The mental disturbance, Doctor Gutzmann argues, results from the embarrassment produced by the peripheral disturbance; but to this view he himself seems to dissent in some measure, for he admits that the emotional disturbance "may be primary." He realizes the importance of the mental phenomena, but chiefly as a result of the peripheral disorder. Ernest Tompkins in this country agrees with a one time associate of Dr. Gutzmann, Dr. A. Liebmann, whom Mr. Tompkins states is of the opinion that the stammering results from an essentially accidental transference of the talking from the subconscious to the conscious realm of mental control; that wrongness of this voluntary control serves to disturb the action of the peripheral mechanism of speech, and thus, through repetition and habit, to induce variably severe spasmodic holdings. The emotional phenomena are also, to the minds of these authors, secondary and due to the embarrassments produced by the peripheral disturbance.

On the other hand, Dr. C. S. Bluemel believes that the origin of stammering is central and is concerned with imperfect functioning of the auditory word center. Through the disturbance within this center the stammerer finds himself unable to visualize rightly certain sounds of the word. This produces hesitation and induces the peripheral spasmodic tensions.

The writer of this article has attempted to prove that the basal phenomenon of stammering is an emotional disturbance. Its origin is usually dependent, he believes, on the emotional instability, together with the peripheral muscular uncertainty in the young child. Emotional shock, fear, etc., serves to induce an emotional upset so great at times as to cause a prompt onset of the disturbance, both in children and (occasionally) in adults. The recent war brought out the importance of shock in this connection. "Social emotion" (emotion induced by social inter-relations) is a highly important mental disturbance in non-stammerers, as well as in stammerers. Stammering, the writer believes, is an especial manner of reaction to this disturbance. The emotional disorder of the stammerer is exactly the "social emotion" experienced by the non-stammerer, and this emotion very largely determines the presence or absence of the stammering. For illustration, a most enlightening fact is the absence, or practical absence, of speech disturbance when the stammerer talks or reads while alone, or to animals, or to young children. Only the "social emotion" induced by the presence of adults is capable of arousing the peripheral perturbations and tensions. Moreover, the presence and degree is also directly dependent on the social circumstances and on the consequent state of emotional excitement of the sufferer. The greatest bugbear of most young stammerers is reciting in school, when speech may become practically impossible. And the stammering once induced increases the social emotion. The "vicious circle" consists of (1) emotion, (2) inducement of stammering, (3) more intense emotion, and (4) more severe stammering.

Behind the individual are in different stammerers an unusual degree of nervous excitability, of rapidity of speech, of tendency to muscular hypertension, etc.,* each forming a background of influence tending to encourage the onset of the disorder. Also as a background of encouragement of the disturbance is severe, or prolonged, ill health. The acute infectious diseases of childhood, as well as prolonged ill health from other causes, often are accompanied, or followed, by stammering. Doctor Gutzmann lays stress on the importance of adenoids, or even of greatly enlarged faucial tonsils. The hindrance to the action of the peripheral speech organs of these essentially foreign bodies makes such a relationship to stammering possible; but the enormous preponderance of non-stammerers among the sufferers from adenoids and hypertrophied tonsils serves to show their relative lack of importance in this relationship, and Liebmann notes that the stammering does not often cease after the tonsil-adenoid operation. This observation the present writer can confirm. It is to be noted also that the emotional shock induced by the tonsil-adenoid operation is capable of producing an increase in the stammering when present, and even of inducing it where it had not previously existed.

Heredity as such seems to have no direct part in the etiology. But inherited characteristics, for example, unusual nervous excitability, may have an important underlying bearing. Stammering, to be sure, is not unusually found among members of the same family. But the son, for example, of a stammering father might stammer by reason of association, as well as through inherited predisposition, or merely by reason of accidental coincidence.

Gutzmann and Liebmann, working together, discovered, through the employment of physical recording apparatus, as well as other experimental methods, that the movements of the chest in speech in stammerers was uniformly less true and certain than in non-stammerers. In 1920 Dr. S. D. Robbins has added another important observation with respect to the physical occurrences in stammerers. He has shown that stammering is accompanied by peripheral vasoconstriction; and also by dilatation within the brain (vaso-dilatation). These phenomena were shown to be also produced by shock and emotion generally. Thus it would seem that emotion may possibly induce disturbance in talking both through its peripheral and its central physical phenomena. While Doctor Robbins lays stress on the effect of the cerebral vaso-dilatation of emotion in clouding the action of the auditory word memory (as previously suggested by Doctor Bluemel), one must also not overlook the possible clouding of the action of the brain in general, of the motor speech directing activities in particular, and certainly not the possible effect, through the discovered peripheral circulatory changes, on the musculature and muscle sensations of the peripheral organs themselves.

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* Such a tendency to muscular tension, when present, would fall closest into Doctor Gutzmann's conception of the cause of stammering. But even here the tension is not of itself stammering, and stammering is induced by the emotions engendered by the peripheral uncertainties of movement; and these muscular uncertainties seem to have a background of causal emotion.

Prognosis.—The mother who takes her stammering child to an American physician for advice is in about nine cases out of ten told not to worry but merely to wait, when the disorder will sooner or later spontaneously disappear. If the physician had had proper instruction during his medical course in this and allied subjects, such ignorant and harmful advice would not so often be given. The occasional light, or recent, case may recover spontaneously. The others will slowly but surely grow worse. And since treatment should be entered on at the earliest possible moment, such advice is the more reprehensible. The "guarantee" of the commercial schools for stammering, of a cure of the disorder in a few weeks, is practically always false. Such a disorder, once it becomes deeply involved in the life and habit of the child, requires great patience and persistence for its complete eradication. Occasionally, when the trouble is light and is taken early, eradication is possible in a few weeks, but usually a variable number of months of careful self control is required. Fortunately, if full intelligent coöperation of parents and stammerer can be secured, complete recovery is usually possible. Without successful treatment the outlook for the child is one of life long struggle and suffering. The most difficult cases are those in which the emotional element, or the excitable element, is pronounced, and those in which intelligence is lacking. And prognosis must always take into consideration the variability and uncertainty of human determination, endeavor, and persistence.

Treatment.—Since stammering profoundly affects the development of the character and habits of the child, and since as time goes on, eradication becomes more and more difficult, it is highly important that the disorder should be arrested at the earliest possible moment. In fact prevention of the establishment of the disorder is to be taught and practiced; but if in the particular case this has not been accomplished, treatment should be instituted as early as possible. As time goes on, self-consciousness and self-abasement with reference to the disorder increase, and a healthy natural mental attitude in social inter-relations is gradually replaced by a tendency to social fear and social isolation; while emotionalism due to the disorder, and also the habit feature, increase in intensity. Thus delay in treatment renders the problem not only deeper but more complicated. In proportion as the disorder has, through its existence during a shorter, or longer, period of time upset the nervous morale and thus become less or more deeply involved in the life and habit of the individual, will self control be easier or more difficult. However, since a certain degree of steadfast effort of the will is required in all cases, and since the will may usually be expected to be more effective as the age increases, the disadvantage from the treatment standpoint resulting from increased age may be offset by the greater intelligence and by the increased activity of the will. Such considerations account for success after childhood period has passed, when the problem often is both complicated and trying.

For a disorder that is so dependent on nervous excitability, on emotional susceptibility, on tendency to muscular tension, and on formed habits, but two ways of therapeutic approach are open, first, through improvement in physiologic and psychologic hygiene, and, second, through the inculcation of methods of self-control. The daily life should tend to normality and strength in all the bodily and mental processes. Sleep and exercise are to be given especial consideration.

When a child begins to stammer, even ever so lightly (for at first the disorder usually presents no evident reason for alarm), careful attempt to discover any reason for unusual excitability, or apprehension, or any perversion of bodily functions should be sought, and, if found, an attempt at correction made. In this field closer observation of the beginnings of stammering will at some later period probably help to clarify the incipient exciting causes. For example, disorders of diet, irregularity of sleep, and social excitement, have together in one observed individual child produced stammering, and their correction has stopped the disorder (report of case by H. Gutzmann). One will do well to see that social relations especially simple are insisted on, that sufficient sleep is secured, that no cause of fears exists, that the child is not pestered by older brothers or sisters, is not crowded intellectually, that the food is agreeing, etc. A subtle something is responsible for these early beginnings, even though they may seem to be merely accidental.

Subjective correction of the disorder finds application both (1) through the conscious direction of the peripheral organs of speech, and (2) through direct conscious psychologic self-direction. In young children the measures to be employed must be concrete and simple. In older children and in adults they must still be concrete, but may be more elaborate. They are in all ages the same in principle. In young children the mother's insistence must largely replace the child's lack of will and lack of interest. If the young child is carefully taught an effective, voluntary, method for control of the chest action for the production of speech, and is persistently guided in its use, the outcome in recovery of the stammering is, in the author's experience, certain. Patience, tact, and intelligence, however, are required. Interference with treatment through causes that produce nervousness should be carefully excluded. Others recommend merely slowness in speech, with other general measures. The author believes, however, based on much experience, that the known insecurity in stammerers of the chest action while talking should always be the standpoint from which physical measures of self-control are instituted.

One reason for the disrepute in which the treatment of stammering has fallen is the superficiality of the methods employed. The commercial schools for stammerers are largely responsible, not alone for superficiality of methods, but also for the inculcation of the false idea that recovery is easy and brief of attainment.

In older children in proportion to the age, and in adults, the methods employed by the author, based largely on those taught him by the late Dr. G. Hudson Makuen, involve the inculcation of voluntary control over, so far as possible, the entire peripheral speech mechanism, chest, larynx, and articulation, and the substitution of educated voluntary control of this mechanism for the natural but perverted subconscious control. The method involves the understanding by the stammerer of the mechanics of production of all sounds of the language, and the use of that understanding in talking. This concrete appreciation of how to direct the peripheral mechanism of speech reacts very favorably on the central mechanism of speech control, and also on the emotionalism aroused by the fear of stammering.

Concerning the methods for aiding the stammerer directly to control his emotional perturbation, it may be said that no human being can master such profoundly human disorder at once, or readily. The difficulty is proportionate to the age of the child (the younger the child the less profound the emotional disturbance, as a rule), to his natural excitability, to his susceptibility to social emotion, and to his tendency to severity of muscular tensions. One must aim to inculcate a spirit of independence and of manliness. One must encourage social intercourse, but always with consideration of the individual success in self-control. One must inculcate an understanding of the significance of muscle tension, and how best to make the effort to secure relaxation. One must also instill the idea of general quietude and relative slowness of speech.

These are suggestions. In the young man, or adult, a great variety of ways for concrete approach to these problems are available. In the child, one must fit his means and methods to the intelligence of the individual. Under no circumstances, however, can one hope to change the nature itself. His aim can be only to wear down step by step, by encouragement of patient self-control, the seriousness of these fears and emotions and tensions. One stammerer may be expected to gain self-mastery by much easier means and in a much briefer time than others. The innate conditions vary tremendously. To gain control, let it be said, of the speech, so that the talking may go on under control with little or no stammering, is not a cure of the disorder. Complete eradication occurs only when through persistence in self-control the **tendency** to stammer has wholly disappeared.

THE PATHOLOGICALLY NASAL VOICE

From the standpoint of the nose, normal speech is constituted of two classes of elemental sounds, one *oral* and the other *nasal*. The difference in quality which gives the two classes their names is dependent on nasal resonance. If the vibrating air from the larynx is permitted by the relaxed position of the soft palate to pass freely into the nasal space there to reverberate, the nasal quality results. If, on the other hand, the vibrating air from the larynx is refused, by the active, elevated, position of the soft palate, entrance to the nose, and is compelled to pass out of the mouth, the nasal quality is absent and the sound is necessarily *oral* in quality. But three English sounds are normally nasal, m, n and ng. Since these sounds appear and reappear in a very large percentage of words, the soft palate is compelled to play with remarkable quickness and ease upwards and downwards as the talking goes on. The pathologically nasal voice is dependent on two factors: First, an abnormal blocking of the nasal passages producing an altered quality of nasal resonance, or preventing nasal resonance altogether; or else (second) is due to elevation of the soft palate for the oral sounds. The effect of these abnormalities is to place nasal resonance into normally oral sounds, or oral resonance into normally nasal sounds. Kussmaul expresses the matter by stating that nasalling results "when the nose is open and ought to be closed, or is closed when it ought to be open." By reason of the two classes of sounds, each subject to its own peculiar pathologic disturbance of nasal quality, and by reason of the almost inevitable functional complications involving both classes of nasalling in the same patient, as well as in a certain class of cases accompanying articulative defections, the subject is complicated and by no means always easy to understand.

Nasalling is best divided into three main classes:

1. Rhinolalia clausa, or closed nasalling.
2. Rhinolalia aperta, or open nasalling.
3. Rhinolalia mixta, both open and closed nasalling in the same patient.

Rhinolalia Clausa.—It is evident that if all passage of air out of the nose was prevented by obstruction, either the nasal resonance would be impossible, or else greatly altered in quality. If the obstruction were complete at the palate (total adhesion of the soft palate to the posterior pharyngeal wall) nasal sounds could not be produced and the m sound would become b, the n d, and the ng g. If the obstruction were complete in the naso-pharynx (extremely large adenoids, or tumor, or complete congenital atresia of the posterior nares), a modified naso-pharyngeal, instead of nasal, or, if the occlusion were in the nose itself, a modified nasal, resonance might result. The doubt implied by the word "might" is made necessary by the fact that in certain of such cases the soft palate responds to the nasal sounds as if they were oral and no nasal resonance of any kind then results. More often, however, complications due to functionally imperfect movement of the soft palate for the oral sounds develop, and the picture becomes one of rhinolalia mixta. Nasal occlusion sufficient to noticeably impair the nasal resonance of the nasal sounds must be considerable. The effect is more pronounced the farther posterior the obstruction is located. Occlusion may be due to adenoids, deflection of the nasal septum, enlargements of the turbinate bodies, or tumor, especially polypi.

A purely functional rhinolalia clausa, due to the raising of the soft palate for the nasal sounds, is a rare occurrence. Very occasionally, after removal of adenoids that have practically completely closed the naso-pharynx and so caused the lowering of the soft palate for the nasal sounds to be practically useless, the soft palate may continue constantly to be raised, as well for the nasal as the oral sounds. The m, n, and ng become then, as has been said, b, d and g. Lermoyez speaks of such a functional closure of the soft palate even in breathing.

Rhinolalia Aperta.—In rhinolalia aperta the m, n and ng sounds are alone normally produced. The essential fact upon which the abnormal voice depends is the considerable passage of air into the nose while the oral sounds are being spoken. The degree of impairment of the voice is determined in general by the extent of the abnormal space for the passage of air from pharynx to the naso-pharynx, or nose, and the patency of naso-pharynx and nose themselves. The larger these passages are the more serious is the abnormal alteration of the voice. The sounds most affected are those requiring the greatest closure of the mouth passage for their production, coupled with the tightest closure of the soft palate. The ē, ōō, ū, ĭ, ā, ĕ usually are most severely affected. The ah and aw are usually the least affected. In congenital rhinolalia aperta, especially cleft-palate, certain consonants are not only nasalized but may be produced with wrong articulative positions. In rhinolalia clausa the speech is always understandable, but in rhinolalia aperta it may be almost nonunderstandable, especially in small children and if the consonantal positions be disturbed.

Rhinolalia aperta may be (*a*) organic, or (*b*) functional.

The organic causes are cleft-palate, congenitally short palate, traumatically short palate, paresis of the palate and partial adhesion of the soft palate to the pharyngeal wall. If the palatal malformation of cleft-palate are operatively closed before the onset of speech, not only is the voice likely to be developed with the least possible abnormal resonance, but the articulative positions and the general accuracy of the speech are both likely to approach close to normal. If the speech be developed with an extensive and broad palatal cleft still open the result may be a speech so little resembling normal speech as to be almost completely non-understandable. Not only are all sounds muddled by the prominence of the nasalling, but the articulative mechanism for the production of many of the consonants may have but little resemblance to the natural mechanism, while the distortions within the word and the elisious at the end of the word may be equivalent to the severest cases of dyslalling. The normally whispered (voiceless, or breath) sounds, owing to the useless escape of so much air through the nose, possess too little carrying power, while the nasality distorts their natural quality. In order to overcome these difficulties the child may produce the p and l (with of course more or less distortion) by a quick explosive sound made in the larynx; and the k by contact of the base of the tongue with the posterior pharyngeal wall. The ng may become an n. The g may become d, or n, or a pharyngeal k. The friction sounds, f, v, both th sounds, s, z, ch, j, all are likely to suffer severely (if produced at all) by their nasality, or by some manner of substitution. Any manner of sigmatismus (malproduction of the s) may be present, and always with intensification due to the nasalling.

Such marked imperfections of the resonance as well as of the articulative positions may occur even in cases operated before the beginning of speech, and especially if the palatal closure still leaves a broad opening into the nose, but their severity is not so great as it would have been if the operation had been delayed, and on the whole the speech results are relatively good, if not perfect. In many cases operated early, where the operative result is excellent, the speech may develop with little nasality and no articulative imperfections.

In congenitally short palate the speech results may be bad, or good, and they depend on the distance of the shortened palate to the posterior pharyngeal wall on phonation, and this varies greatly. These cases are less well known and less common than those of cleft-palate. They represent, apparently always, a process of congenitally healed cleft-palate, since a track can be outlined beneath the mucous membrane along the central line of the hard palate, and the latter may be ended posteriorly by a triangular indentation at its center. The soft palate is the part particularly affected by shortening. Such cases sometimes are seen in families where true cleft-palate has occurred and in certain of the feebleminded.

Following tonsillectomy, if the posterior pillars have been injured severely, and especially if the soft palate itself was relatively short before operation, the contractions of the scar tissue, in connection with the injured pillars, may so tense the palate as a whole that it is no longer long enough to perfectly fulfill its function in speech, of closing off the naso-pharyngeal space for the oral sounds. Rhinolalia aperta is then the permanent result. The author has studied five or six such cases. They emphasize the need of accuracy and delicacy in performing the tonsil operation.

Rhinolalia aperta functionalis occurs in children who by reason of prolonged illness, or of feeblemindedness, have never acquired the function of raising the palate for speech purposes, or have acquired the function imperfectly. It occurs also in children following adenectomy in cases in which the naso-pharynx has been nearly closed by very large adenoids. The adenoids have so nearly taken the place of the palatal function, that the palate has habituated itself to inaction. And functional rhinolalia aperta occurs also following recovery from paresis of the soft palate due to diphtheria. A habit and a false appreciation of the sound qualities has been formed during an unusually prolonged period of inaction due to paresis, and consequently inaction of the palate continues after the paresis has recovered.

Rhinolalia Mixta.—The organic conditions which produce the combined disorder are relatively common. They are nasal occlusion, or naso-pharyngeal occlusion (adenoids), coupled with cleft-palate, or short palate. Especially if the palatal defect be pronounced the rhinolalia clausa is an advantage. For the conditions which produce rhinolalia clausa need be removed only to any degree desirable, and the seriousness of the rhinolalia aperta is mitigated by the existence and continuance of the obstruction which produces the rhinolalia clausa. A moderate degree of functional rhinolalia aperta is not uncommon as a complication of rhinolalia clausa. The reason is essentially psychologic, being dependent on the local sensational, and the auditory, confusions engendered by the nasal, or naso-pharyngeal, obstruction.

Diagnosis.—In marked simple cases of either closed, or open, nasalling the diagnosis is easy. In the lighter simple cases, especially if functional, a clear understanding of the character of the voice defect may require experience and study. In the mixed cases the differential diagnosis may be difficult. To distinguish between open and closed nasalling the following method, long described in the German literature, if executed correctly, is delicately determinative. Ask the patient to say the sounds "ah-ē" distinctly. Then occlude the end of the patient's nose with the thumb and finger, and have the same sounds repeated, cautioning the patient to speak clearly *out of the mouth.* In rhinolalia clausa no change in the nasality should occur due to closing the nose, but in rhinolalia aperta the nasality, especially of the ē, becomes more pronounced. In this test the danger is that the patient, because of the artificial nasal holding, may *produce* a careless functional rhinolalia aperta. If doubt still exists one may pass a diagnostic rubber tube, arranged with suitable hard rubber tips, between the patient's nose and his own ear, and, using suitable sound combinations, note which combinations, those containing nasal sounds, or those containing only oral sounds, are nasal. If the oral sounds are nasal the trouble is rhinolalia aperta; if the nasal containing combinations alone are nasal the disorder is rhinolalia clausa. Physicians too frequently err by fixing their attention in certain cases wholly on closed nasalling, and serious errors in operating have resulted from this mistake. Open nasalling is by no means uncommon.

Prognosis.—Modern surgery makes it possible to remove any manner of nasal and naso-pharyngeal obstruction, at any rate sufficiently for speech purposes, excepting possibly extensive adhesions of the soft palate to the posterior pharyngeal wall. With this latter cause sometimes excepted, the prognosis in rhinolalia clausa is excellent. In cleft-palate prophylaxis is of great importance. It is secured through operating before speech has begun, and by instituting speech training therapy as early as possible after operation; and, in any case, by affording the child the best possible examples of speech. In operated cases and in congenital short palate the prognosis is dependent on the distance between the soft palate and the posterior pharyngeal wall as compared with the length of the soft palate, and its capability of movement; also on the degree of movement of the pharyngeal musculature (especially the superior constrictor) on phonation; and, finally, on the relative narrowness of the naso-pharyngeal-nasal passages. The combination of operation with adequate speech training should serve to banish articulative malpositions; in the most favorable cases afford a fine voice and speech; and in all others an understandable, albeit still more or less nasal, speech. Operating is often of little speech value without the after speech training. Through training methods purely functional cases are all correctable in intelligent patients.

Treatment.—For simple organic rhinolalia clausa operative removal of a sufficient amount of the occlusion is curative. Before operation is undertaken the surgeon should be certain that rhinolalia aperta has been excluded. If nasalling continues after reasonable removal of obstruction, the preliminary exclusion of rhinolalia aperta was wrongly determined. In congenital cleft-palate the time of choice for operative closure of the cleft is before speech has begun, and the second choice of time is at about five years with the understanding that speech therapy is to follow the operation immediately. In either cleft-palate, or congenital short palate, an obturator is seldom advisable in children, although sometimes of great value in adults.

Gutzmann advises in selected cases the injection of paraffin beneath the mucous membrane of the posterior pharyngeal wall, in such a manner as to narrow the passage from the oro- to the naso-pharynx. This procedure is undoubtedly valuable from a purely mechanico-acoustic standpoint, but in cases of paraffin injection for other purposes so much later trouble due to displacement of the paraffin and to sloughing has resulted, that one needs to be very cautious in deciding on such a procedure. It is to be noted also that such treatment does not usually make training unnecessary.

One principle having reference to operative procedure in organic rhinolalia aperta, about which certainly the rhinologists of the United States seem almost universally unaware, deserves positive emphasis. This is the principle already mentioned, that the nasality of organic rhinolalia aperta is improved just in proportion as the naso-pharyngeal-nasal breathing space is small. Therefore the disposition of rhinologists to render the nasal breathing passages in rhinolalia aperta normal (as might be desirable if no question of voice entered) needs emphatic correction. In this condition removal of any obstruction, either of the nose or the naso-pharynx, which is not rendered positively necessary for some important reason, as, for example, disease of the middle ear, is bad rhinologic practice, and results often in positive and permanent harm to the patient. Gutzmann advises that adenoids, if necessary to be operated either for rhinolalia clausa or for middle-ear inflammation, should be removed only in part, as much of the mass being left as careful judgment permits. Not only is this good practice, but enlarged posterior ends of the lower turbinates (vasomotor enlargement of the turbinate bodies), and deflections of the septum, in the absence of clear production of harm, are to be considered in this connection as positive blessings. If for reasons of the closed nasal voice, or in order to render nasal breathing possible, or for other reason, it becomes necessary to operate for nasal occlusion, the operation should aim to open the breathing space only in so far as is necessary for reasonable accomplishment of the object sought and no further. At the very moment of writing the author has been consulted about the speech in a cleft-palate patient, who two weeks previously had been operated by a rhinologist for adenoids and tonsils in the thorough manner above warned against, to the positive and permanent harm of the patient.

The principles of speech therapy in organic rhinolalia aperta and congenital short palate are based on the following facts: First, the strong nasality has dominated the sound pictures and the mechanics of speech production so far that the child has usually acquired wrong habits of speech production, in the direction of a greater nasality than is compelled by the organic defects; second, the waste of air through the nose while talking has not been overcome by a more energetic use of the chest, as compared with its use normally, in talking; and third, the sound confusions due to the strong nasality have often led to wrong methods of producing the elemental sounds and their combinations.

For correction of wrongly produced elemental sound positions one proceeds as with similar defects in other classes of patients. The nose for this purpose may be held closed momentarily while practicing on individual positions, in order to obtain a full stream of air out of the mouth. With proper persistence, coöperation, and skill, success in gaining normal articulations is to be expected.

In practically all the training work attempted in open nasalling the basis of effort should be in the chest. The patient should be taught to talk with the chest well filled with air, and to produce the act of expiration for speech purposes with controlled, increased energy. It is difficult to attain to an easy, natural, and yet energetic management of the outgoing breath even in older children, and especially so in younger children. In proportion, however, as the production of the voice rests on chest action and chest resonance, will the nasality become less objectionable. In the most favorable cases, if the patient is intelligent and persistent, an excellent voice resonance can be attained.

Improvement in the nasality is directly approached by training the child to throw his efforts into expulsion of air in talking out of the mouth, rather than out of the nose; by encouraging openness of mouth in production of speech in so far as possible; by encouragement of depression of the back of the tongue (free passage of air out of the mouth) in the production of certain vowel sounds; by education of the ear to clearly appreciate the difference between nasal and oral voice resonance; and, if possible, by encouragement of the closure action of the superior constrictor muscles while talking.

Where conditions are favorable the employment of an instrument like Gutzmann's hand obturator is highly helpful. The instrument has a sufficiently long steel handle to be held by the instructor; its distal end is so constructed as to be suited either to raise up the palate, or to act as an obturator between the oro- and naso-pharynx. The practice alternates first with, and then without, its use. It serves chiefly to educate the patient's ear to realize when his voice is, and is not, nasal. A soft rubber tube with suitable tips for insertion at one end into the nose and at the other into the ear may with older children be extremely helpful in enabling them to measure their degree of success in eliminating the nasalling. Functional rhinolalia aperta, or rhinolalia clausa, is treated on the principles above laid down.

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CHAPTER CLXXXVII

THE SEXUAL LIFE OF THE CHILD

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INTRODUCTION

The sexual life of the child is of the utmost importance to medical interests, first because of its significance in the child's racial and social usefulness, and secondly because of its inseparable relation to the entire somatic and psychical life of the individual. It is essentially a psychical rather than a physical problem, but because of these factors, it becomes one in which the two points of view are inseparably fused.

The genetic concept, therefore, both physically and psychically considered, makes its strongest appeal in this department of child study. Confronted with the vitally dynamic nature of the material with which the physician has to deal here, the wonder is that we in our medico-psychological thought could have been so long at a standstill. For at a standstill we have been, or at best, moving about in a bewildering circle. As physicians, we have not understood how to get below the purely descriptive occupation with this subject, although the child world has stood beckoning into depths of peculiar richness and possibility. Nor has it helped that each one of us has passed through the childhood period not so very long ago. The darkness of understanding and the actual fearfulness with which the adult ordinarily goes back to the sexual aspect of his childhood has led him for the most part to deny its existence. Its reality and vitality have retreated past memory and imagination and thus past criticism and judgment. In fact, we fail to understand the entire psychology of the child, which is so intricately interwoven with his sexuality that the two are inseparable. The inability, therefore, to understand the former has led to the failure to admit the richness and actuality of the latter.

For psychology and medicine also have been seriously blinded in their approach to this subject. They have denied themselves actual entrance into the problems existing there, and have confined themselves to partial understanding of symptoms, to limited diagnosis, and to ineffectual treatment. Psychology has remained unequipped to give that wider advice sought by parents for the aid of their children in the peculiar problems confronting each life from its conception to its maturity. And medical practice has held itself aloof generally from entering into the broad psychical background of disease. It has therefore failed to understand the more conspicuously psychical nature of many childish disorders, which manifests itself either in slight abnormalities of development or adaptation, or in actual functional and perhaps organic diseases, all of which are caused largely by the child's difficulty in adapting himself to the conditions of life as he finds them and by his failure to attain their mastery.

Psychology has been as timid as the parents themselves in admitting the child life to a share in this full genetic process of uninterrupted growth from the moment of conception of a sentient being to the age of puberty. It is only at the latter period that the practical and the intellectual world have first ascribed to the child certain impulses, an onslaught of feeling, and an awakening to need for adjustment. Curiously oblivious of his own childhood, the adult fails to understand that were such new powers suddenly called into play at a time when the child is supposedly unprepared for them, they would, without doubt, completely overwhelm him.

This misunderstanding has been most marked in the matter of sex, towards which the attitude of the adult has been marked by a curious aloofness. The average child has not, on this account, received the sympathetic appreciation and instruction which would not only help him in his preparation for puberty to a fuller understanding of the disturbing phenomena of that period but which would also enable him to adapt himself more effectually to his surroundings. Understanding of the sex phenomena of childhood would help to set in their rightful place the many things which have been a perplexity to the psychologist and medical attendant and a source of bewilderment and anxiety to the parent, and would aid in the effectual treatment and correction of certain anomalies of conduct and psychical development and the medical disturbances arising from them.

The individual at puberty is not infused with new powers. There are no latent forces set aside by nature to be overwhelmingly awakened when the child has reached a certain stage of development. If it were once realized that there are no distinct internodes in the process of growth, the child would be far more completely set on his way to becoming a well-adapted citizen of the world, prepared to meet and appropriate to his best interest and greatest productiveness whatever the world has to offer.

Nature has a great regard for unity. Violent eruptions, either in the process of construction or destruction, have no place in her economy. The forces at work have been slowly preparing, whether or not we are enabled to see their movement. Such is the conception that has made its way in recent years into psychology and medicine. This concept is termed dynamic unity. It expresses itself not only in the average course of development of individual life in all its phases, but also, when development is retarded or complicated, in disease and variability. Disorders of body or mind, therefore, of whatever degree or variety, are only so many diverse expressions of the same dynamic force or effort expressing itself thus imperfectly. For this reason, normal sexual development and various sexual anomalies are to be considered a part of this dynamic self expression and dealt with accordingly.

This form of approach to human problems, above all to the most essential problem of child development, makes it necessary to adopt the notion of this unity in human nature and to utilize the hypothesis of the preservation of past psychical life as the basis of such unity. This preservation of past experience is the source from which stream continually the elements which infuse their psychical color and meaning into every new experience with the outer world. It alone gives a genetic unity and completeness to psychosexual as to all development.

We accept the concept of the unconscious to designate this preservation of past experience and its continuity and activity in relation to the superficial life which is manifest daily before our eyes. It must be remembered in doing this that, in the unity of nature, the child is one of an unbroken race. He has condensed in the brief period of embryonic existence the millions of years which have brought evolution from the free swimming cell to the complex physical organization of man. He has passed from the rudimentary primeval sensation of the amoeba, which responds with a slight chemical acceptance or rejection to a particle in its surroundings, and which is vaguely sentient of an impulse to divide and reproduce its cell, to the most complex refinement of psychic pleasure and pain known to man and the mighty element of intellectual regulation and control.

Embryology has written in unmistakable terms the hurried recapitulation in one small individual of the immeasurable period of physical evolution. Traces of this evolution still exist in the inheritance of organs which were once useful but are now no longer needed directly and which still persist with those later acquired and adapted for use.

Nor does the child's recapitulation stop here. He has still in the earliest years of his conscious life to make, also in a condensed form, the same journey of trial and error with which early man, the child's own primitive ancestors, brought the manifold and often stupendous elements of his environment to the touchstone of his experience, with which he not only tested and gained slow mastery of his environment, but also, through his changing reactions, built and modified his own experience.

All living forms are a vast record of this preservation of what is useful in their past inheritance. Geology and paleontology bear excellent testimony to the imperishableness of history and to the fact that much that is laid away because its service is only indirectly required is not therefore discarded. In the human psyche, the final product of this long evolution of sensation and feeling, developing its highest form in the human intellect, there still are relics, traces of former activity and serviceableness. No longer suited to present conditions they are still active as a part of living nature in some form, and constitute living forces to be understood and taken into account. They are like the non-living records of past ages which constitute and support the earth's surface and which serve to record and interpret the history of the past. In just so much, moreover, as they are vital and functioning, must they be controlled and directed to a higher form of activity suitable to present-day life and governable by present-day culture and need.

Such is the background of the sexual life of the child. Such are the elements of its past inheritance, which demand understanding and watching, and which are to be guided properly along the path of development. The brief recapitulation of the experience of the race which the child practices in the early years of life should prepare him for his environment and direct his forces so that they shall aid in the creative immortality of the race. This heritage makes him sexually a richly dynamic creature, containing not only the germ but the greater psychosexual setting of the physical immortality of the race, which in its subsequent development infuses and energizes that creative power within him upon which progress in civilization and culture depends.

THE GENITAL ORGANS. THE STRUCTURAL BASIS OF THE SOMATIC SEXUAL LIFE

A study of the sexual life of the child in order to be illuminating and practical must therefore be based upon the dynamic unity of the individual life. This is expressed in the concept of an energy distribution and transposition which permits of self-expression of the inherent life force upon varying levels of activity. Accordingly it is more complete and more comprehensible to speak of the psychosexual life of the child. This unifying energy concept however establishes the functional unity of mind and body and permits us to conceive of the physical organism as primarily and simply the means of expression of mental activity. Structurally, apparently, it has been developed by slow degrees to meet this psychical need for such self-expression. The organism is not concerned solely with conscious self-expression, as we have been accustomed to think of the self, but it was for ages exclusively, and is now still in large part, at the service of unconscious impulses and power.

Definiteness of structure and perfection of adaptation are nowhere more beautifully shown than in the human sexual organs and their highly organized physiological and physiological-psychical functioning. Hence a brief description of the anatomical apparatus and of this functioning is necessary before one can discuss the broader psychosexual life and all that it involves in the child's development.

The sexual organs of the male consist of the penis, the anterior portion of which is visible externally. Behind this hangs the scrotum or sac containing the testicles or testes and their accessory organs. The testicles are two ovoid structures, each suspended in the scrotum by its spermatic cord. Each is surrounded by three tunics: A serous sac, the tunica vaginalis; a strong fibrous sac, the tunica albuginea; and a vascular tunic composed chiefly of blood-vessels held together by areolar tissue. This sac is known as the tunica vasculosa or pia mater testis. From the tunica albuginea fibrous septa pass into the interior of the testicle dividing it into a number of separate lobules. These again are composed of seminiferous tubules, which by branching and convolutions greatly increase the area in which the spermatozoa are formed. These seminiferous tubules unite to form the vasa efferentia, the efferent ducts for the transmission of product of the testicles. There are about 12 of these for each testicle. As they pass out of the testicle they are coiled in conic masses to form the epididymis, which is situated at the top of the testicle, into the single tube of which they empty. This tube leaves the epididymis as the *vas deferens* and conveys the testicular secretion to the exterior. It traverses the inguinal canal to the abdominal cavity and passes down to the prostate gland which surrounds the proximal part of the urethra. The latter is a canal running the entire length of the penis. It opens at its proximal extremity into the bladder and serves for the passage of the urine from here to the external opening of the urethra, the meatus which is situated at the anterior extremity of the penis. The prostatic ducts convey the contents of the prostate gland into the urethra. Just behind the prostate, the vas deferens forms on either side a diverticulum known as the seminal vesicle. This constitutes a reservoir for the semen. Its duct joins the vas deferens thus forming the common seminal or ejaculatory duct leading into the prostatic portion of the urethra, by which the exterior is reached. Behind the posterior portion of the urethra there are also the glands of Cowper, or suburethral glands, which likewise discharge their contents into the urethra. Other glands are found in the walls of the seminal vesicles, the vasa deferentia and the urethra.

The penis consists of a root attached to the descending portions of the pubic bone, the body or main substance of which is made up of three cavernous portions, the paired corpora cavernosa and the corpus spongiosum. They consist of a spongy mass known as erectile tissue, with lacunar spaces within which a quantity of blood can be retained. The urethra perforates this tissue. The terminal portion of the penis, which is known as the glans, is covered by mucous membrane and ensheathed by the prepuce or foreskin.

The female genitalia are no less adaptively constructed than the male for the function they have to perform in reproduction. Externally they constitute the vulva, which consists first of two large folds known as the labia majora, situated on either side of the median line. Two smaller folds, the labia minora or nymphæ, lie just within these. These serve to enclose two orifices in the median line. The anterior one is the meatus or urethral orifice, which forms the exterior aperture of the urethra, which in the female is a short, almost straight tube leading from the bladder. The posterior aperture is the opening into the vagina, which is partially closed by a portion of mucous membrane, varying in form and extent, but usually of sufficient size to obstruct entrance to the vagina until it has been ruptured. This is known as the hymen.

The labia minora diverge posteriorly to admit the vaginal opening and converge anteriorly to meet at the clitoris. This organ is the analogue of the male penis, and is of much the same structure, though without the canal. It consists of erectile corpora cavernosa and an extremity known as the glans. Anterior to the clitoris and in front of the pubic symphosis is a rounded eminence, which in the adult is covered thickly with hair, known as the mons veneris. Posterior to the clitoris, beneath the mucous membrane at either side, there is a further mass of erectile tissue known as the vaginal bulb, or bulb of the vestibule. Corresponding to Cowper's glands in the male, are Bartholin's glands, whose ducts have their orifices on either side of the vaginal opening.

The interior of the vagina is tubular and flattened so that during quiescence the anterior and posterior walls are in contact. The posterior wall is longer than the anterior wall and thus reaches up higher about the cervix uteri, which extends down to meet the vagina. The uterus is a muscular pear-shaped organ consisting of a broad flat portion above and the elongated cervix below. At its upper end there are two lateral apertures, the orifices of the Fallopian tubes or oviducts, which connect the uterus with the ovaries.

The ovaries are the female glands of reproduction corresponding to the testicles in the male. They are somewhat ovoid in shape and are situated one on each side of the uterus, at its upper extremity. They are lined by peritoneum and consist of a layer of fibrous tissue, the tunica albuginea, and a mass of vascular connective tissue. This contains a large number of vesicular structures, the ovarian or Graafian follicles, each one of which contains an ovum. During the years of female reproductive activity, one of these follicles ripens normally every four weeks, bursts, and expels the ovum, which passes through the Fallopian tube into the uterus. If it is fertilized by the male spermatozoon, it remains here to develop into an embryo. If it is unfertilized, it is soon cast off from the body.

At regular intervals of four weeks there is an increased flow of blood to the uterus, and a congestion of the ovaries and external organs, with gradual outflow through the uterus and vagina to the exterior. This process is known as menstruation. Its physiological relationship to ovulation and pregnancy is not clearly established, but the alteration of the mucosa which accompanies it suggests that it is a preparation for the nidification of the fertilized ovum. It ceases normally during pregnancy, while its appearance at puberty and its complete cessation at the menopause are practically coincident with the term of female reproductive activity.

In the child of either sex the genitals differ from those of the adult mainly in size and in lack of functional secretions. The most conspicuous difference is the absence of hair which in the adult male surrounds the root of the penis and in the adult female covers the external genitalia.

The changes which puberty brings about in the male organs are noticed also in the greater size of the penis due to increase in the corpus spongiosus and corpora cavernosa. The prepuce becomes more retracted and more freely movable upon the glans. There is an increase in the sensitiveness of the organ. The testicular glands increase in size and develop in function. The scrotum enlarges and the testicles, which normally descend slowly from the abdomen toward the scrotum during embryonic life, descend still further as they enlarge, while the spermatic cord lengthens. The most important functional change which takes place is the formation of spermatozoa, the active reproductive element, as well as of the inner secretions.

The enlargement of the breasts is one of the most conspicuous changes in the female at puberty. There is also a noticeable difference in the shape of the external genitals. During childhood the vulva is more distinctly visible, being placed higher and toward the front, while the labia majora are marked by the absence of the fat which is deposited in them at the approach of maturity. Important changes also take place in the development of the clitoris at puberty. This organ increases in length, and there is a corresponding increase in the size of the terminal nerves supplying the erectile tissue. The uterus, the growth of which was inhibited during childhood, develops rapidly during the period of puberty. The regular occurrence of menstruation leads to changes in the mucous membrane, while ovulation also greatly alters the appearance of the ovaries.

This briefly is the anatomical structure which is to subserve the most important function racially of the human organism. Psychically, therefore, as well as physiologically, it attains an importance even in the child's life which has been too long overlooked, or relegated to a later period of education and care. By this time however the psychosexual life is already well on its way toward a permanent establishment, which will mould and color all the future life. Physiologically the child is functionally immature, but just as the anatomical structures are developing toward their adult form and size, so too the functional activity is gradually establishing itself, and already both form and functional stimulus are rousing in the child an active and normal curiosity and interest. They therefore have an immeasurably important bearing upon his later life, which cannot remain unrecognized and ignored.

A brief review of the normal sexual function will serve as a basis for understanding its beginnings in child life. The sperma are located in the basement membrane of the testicles, where in early life they pass through the different stages of development, until they become the mature, motile spermatozoa. These spermatozoa are present in enormous numbers in the seminal fluid, which is composed of secretions from the testicles, prostate, seminal vesicles, and Cowper's glands. There is evidence also pointing to the fact that there is an internal testicular secretion which enters the general circulation and plays some rôle in the development of the secondary sexual characteristics of the male.

The ovum is ripened in the female ovary by a process similar to that by which the spermatozoa are formed. Situated in the cortex of the ovary are the Graafian follicles in which the matured ovum is found and from which it bursts to pursue its way into the oviduct. The formation of these Graafian vesicles is begun during embryonic life, but it is not until puberty that they develop in size and abundance, from which period, normally, their functioning is maintained uninterrupted until the menopause. The rupture of one of these follicles, which occurs at each ovulation, leaves a cicatricial scar.

The sexual impulse and the sexual act which follows it are the means by which the sexual products are brought into the service of reproduction. The physiological and psychical processes of this function can never be completely separated. Together they have gone through a process of development which is maintained throughout the entire period of childhood Thus the child cannot be fully understood without a knowledge of the functioning of the psychosexual process—and the process itself cannot be understood without a knowledge of its development during the life of the child.

The normal sexual act presents a series of stages in a definitely combined physiological and psychical process in which there is a close interrelation of the two forms of stimulus. The terms of nervous activity involved correspond to the latest researches in the field of vegetative neurology. The process consists of four distinct phases. The first may be designated the phase of desire, or libido, in the restricted sense. The second stage is that of erection, the third that of ejaculation, and finally the orgasm. These are most conscious in the active functioning of the male, and have their precise homologies in the female.

The stimulation of desire is dependent upon external psychical stimuli, such as arise from various forms of contact with the love object, sight, smell, touch, everything, in fact, that appeals through the avenues of the senses. Memory also plays an important part in arousing desire, serving to recall such sense appeal and producing a phantasy stimulus, especially in the cases where the normal impulse tends to become even slightly pathological. These psychical stimuli become active so far as the genital function is concerned only through a definite mechanism, which in turn serves to arouse and increase the psychical influence.

Higier quotes Müller-Dahl as saying that the higher cerebral activity is made more susceptible to sensuous impressions and erotic ideas by the products of internal secretion of the sexual glands. Where these products are wanting, as in the child or eunuch, whatever partial feeling may exist, there is no complete desire sufficient for the performance of the sexual act.

Certain inner secretions of the genital glands, known as hormones, entering the blood current, influence the desire. In the male, there are produced probably from the interstitial testicular substance and appear after birth coincident with the semen.

In the female, in whom sex desire is increased with menstruation, they are probably produced from the interfollicular cells in the ovary. The hormones act as connecting link thus between the somatic division of the organism and the reproductive portion.

The different stages of the sexual function are not controlled by any special cortical center, but through the sympathetic and autonomic innervation. The secretion both of the hormones and of the excretory product in the male is called actively into play, chiefly by cerebral activity aroused by psychic stimuli in the form of the sense impressions, memories, or voluptuous ideas. The cerebrospinal fibers pass through the spinal cord at the upper lumbar segment, which is the erectile synapse. The sex impulses stimulate erection, which is a vasodilator phenomenon. The blood supply to the penis is increased, filling the lacunar spaces of the spongy erectile tissue and causing the penis to increase in length and thickness and to become hard and inflexible. Erection is also induced through a periphero-spinal reflex in which the stimulus arises in the skin or mucous membrane and is carried through the sympathetic ganglia to the second sacral segment, which then sends its impulse to the periphery. There is also an autonomic reflex which induces erection, probably located in the ganglia cells of the hypogastric plexus. This is activated by the fulness of the seminal vesicles or bladder.

The third stage of sexual activity is that of ejaculation, in which both the spinal and sympathetic systems are concerned. The sympathetic reflexes are further stimulated as a result of contact movements acting upon the erect and excited penis, which induce, through a summation of stimuli, a peristaltic muscular contraction of the vasa deferentia, seminal vesicles, and prostate. The reflex response is the production of the orgasm with the emptying of the mixed secretions into the prostatic portion of the urethra. Immediately upon this occurs a muscular contraction of spinal reflex origin, by which the semen is finally ejaculated. The relaxation of the penis which follows depends partly upon the passive relaxation of tone in the efferent nerves, and partly upon the active contraction of the smooth musculature of the skin of the organ and of the corpora cavernosa and corpus spongiosum.

The sexual function in the female bears a close analogy to that in the male, though the female, usually by repression, assumes a more passive rôle. The clitoris in the female child, which is the homologue of the penis in the male, forms the chief erogenous zone, but its excitability must at puberty undergo a repression and a displacement to the vaginal region.

This is often accomplished only after a certain amount of sexual experience. The clitoris remains, however, to a limited extent, actively responsive to the erectile stimuli, and thus participates in the initial stage of excitement. The main stimulus activity, however, is induced by contact through the vagina, and is accompanied by the same psychic influences which are active in the male. The orgasm is also produced, normally followed likewise by an ejaculation, not of the reproductive product as in the male, but of certain secretions from the Bartholin's glands and the other mucous glands of the uterus, vagina, and vulva. Even at the beginning of the sexual act the genital passage is prepared by a certain amount of secretion from local glands.

In both male and female the sexual act is accompanied by increased activity of internal organs, such as the heart and thyroid, by increase of respiration, and by certain characteristic active muscular movements. With the relaxation occurring in connection with ejaculation, following upon the acme of excitement and activity known as the orgasm, there follows a general relaxation and pleasurable sense of fatigue, psychical as well as physical.

This is the usual process prepared in the economy of nature for the most fundamental and necessary racial function. Nature, therefore, has established it upon a delicate and complexly adjusted mechanism, admirably adapted to its fundamental biological purpose, and has at the same time fortified it by associating it with the most intense of pleasurable sensations and by surrounding it with all that gives value and stimulus to life in the widest sense. In this way the sexual life has been established and conserved, not only as the unfailing source of creative life, but also as the basis of endless extension and elaboration into all forms of effort.

It is from this latter point of view that the sexual life of the child acquires such significance and becomes so vital a factor in the consideration of the parent, the educator, or the physician. Any person whose task is the guidance and training of youth should understand the sexuality of childhood thoroughly, not only in its usual aspects but, what is even more important, in such deviations from the healthy as occur in childhood and continue their influence on the future adult. He must likewise appreciate the fact that the psychosexual development threads its way through the whole fabric of early existence, causing many of the anomalies of growth and certain ailments of childhood, all of which, without this recognition, are difficult of explanation and treatment.

The sexual organs and their functioning in the sexual act have been briefly outlined only in their strictly healthy condition. The deviations from this are countless, ranging from the slightest individual peculiarity of structure or function to the extremes of pathological development, inability, or perversity. These, however, are functionally the result of a disturbance in early development and of fixation upon one of the various stages through which the psychosexual development proceeds. Morphological anomalies are of significance to the child only in so far as they disturb his gradual psychosexual development and fix attention, through weakness or other variation, upon the genital area itself, or upon the phenomena which precede and are allied to adult sexual activity, such as the excretory functions. The resulting disturbances arise early in development, and are perhaps either organically conditioned in embryonic life or due later to fixation upon the various stages through which the psychosexual unfolding of the child must make its way. This matter, however, will be discussed in its place in the direct consideration of the sexual life of the child.

SEXUAL DEVELOPMENT THROUGH THE VARIOUS PERIODS OF CHILDHOOD

What makes up the sexual life of the child? In considering it, are we merely investigating a portion of his equipment, separated by structure and use from the rest of his organism, distinctly set apart to be used only for a definite period of his later life? Or, in dealing with the subject as it affects the problems of medicine, do we merely concern ourselves with certain variations which may manifest themselves and which mark certain individuals as physiologically perverted in development and unusual or even viciously anomalous in behaviour? Either one of these points of view separates the period of childhood from the unity of evolutionary and genetic growth and depreciates the importance and influence of the sexual life in the economy of each individual development.

The normal genital development, with its production of the inner secretions, has a continuous effect on growth and differentiation, so that we cannot separate it from the welfare and the life history of the entire organism. Before, however, we can establish the intimate correlation, we must first disabuse the mind of certain inhibitive forms of expression which have relegated this subject to the more striking anomalous manifestations or have regarded as abnormal that which rightfully belongs to the child as a part of his natural development and which has received recognition as a phase of normal sexual life only after the period of puberty. Above all, it is necessary that the physician should comprehend more exactly the place sexuality occupies in early child life.

A definition of sexual life at once inclusive and essentially fundamental suggests the reason of its importance in childhood. In its widest sense it is the effort for self-expression, which has been established in the very nature of evolution as reproductive and creative. In its narrower sense it is related to the basic biological means and manner of functioning, to the process that makes reproduction a definite racial fact and conserves the creative function in individuals also endowed and equipped for its wider exercise. The child, therefore, enters the world bearing within him an inherent force which is both active and receptive toward the opportunity which his environment puts within his reach. He has the sensory acquirements, both those belonging to his race and his species, and those inherited by evolution through the species which have preceded his own. He is therefore fitted to receive impressions from his very first contact with a limitless and varied environment. At the same time he contains within himself the fundamental racial force, made individual for him, by which these impressions are to be appropriated and utilized. It urges him to his end in life—to impress himself upon the external world, to express himself through the external world. This goal, because of this insatiable energy, this life force, must of necessity be a creative one.

It is not possible, as psychologists have attempted to do in the past, to examine one by one the distinct sensations which a child experiences, and the response he makes to each, and thus gain a clear working knowledge of the child mind, its impulses, endeavors, its progress in development. This method has succeeded in furnishing some descriptive information in regard to child behavior. It has, however, explained nothing of the dynamic activity of the child mind. It has explained nothing of the emotional and motor responses which make up the complexity of his life and cause the perplexity of those who have that life in their responsibility. Human nature is too much of a vital dynamic whole, it is too evanescent, "new every moment," to be investigated by any such cut and dried method of dissection. The child in his sexual life, as in any other phase, presents no single psychical fact. He cannot be approached as other than a creature of constantly streaming impulses and activities which arise from out his deeper hidden life, and are modified, activated, and actualized by the constant necessity of adaptation to an encircling environment. Nevertheless, some form of analysis is necessary in order to reach an understanding of the problem before us.

Observation may form the basis for such an analysis. The child's activity, such as his speech and his motor acts, his evidences of emotional disturbance or satisfaction, both directly and indirectly manifested, are the material from which knowledge must be gathered. These signs, however, are all barren without some further means of interpretation. Many subtle manifestations have been slighted in the past and more evident ones have remained hidden because there has been no common denominator which would include the most indirect as well as the more direct manifestations and set them all forth as the expressions of an active and fertile mentality. In fact, any relation between such a mental life and the noted physical activities and emotional states was seldom considered. Irritability, moroseness, inattention, ill success at school, thumb sucking, nail biting, urinary disturbances, and various pathological conditions were all explained either by outward physical causes or, at best, as superficial psychic disturbances.

The first step in attaining this fundamental interpretation of the psychosexuality of childhood is to perceive the unity in the constantly shifting movement of psychic development. In this way alone can we obtain the real factors at work in the sexual life of the child, and understand its possible relation to those various manifestations by which he indicates his adaptation or maladaptation to his world. Once this essential and vital unity is grasped, it is then necessary to enter into a study of its components in order to understand the anomalies of childhood and attain, to any extent, control and direction of sexual development.

Ordinary conscious retrospection on the part of the adult is next to useless. The factors which played the largest part in puzzling older investigators were those dealing with things which the children do not willingly reveal to themselves, and which in the older generation, therefore, as far as consciousness goes, have been completely relegated to oblivion. They are in general the things which the child finds difficult or incapable of fitting into his surroundings, or what he conceives to be the adult's conception of his surroundings. This presupposes* already in the child life a preserved past of experience from which his conscious mental processes draw, and a past to which his daily experience is adding. His life is in a state of flux and all the gathered material of his activities, of sensation and associative processes is also sorted and that which is useless to the limited field of consciousness and incompatible with his social position is soon crowded out of consciousness. His mental life becomes on this account increasingly difficult of access to the observer, and yet because it is dynamic the influence of the banished material continues to play upon present and future states and acts. These acts and conditions need then to be watched with a keenly interpretative eye in order to gain insight into psychic material that lies below them.

The child, however, is but repeating the repressive experience through which the adult has gone. The interpretation therefore must be sought by other than direct means. Psychopathology has here come to the aid of the child psychologist. "The hysteric," it has been said, "suffers from reminiscences" and these have been found by repeated clinical investigation to be reminiscences of childhood, and more strikingly still of the sexual life in childhood. These, like the reminiscences of other forms of psychoneurotic disturbances, are those which have not fallen successfully under repression. They manifest themselves in a variety of disguised symptoms, whose disguise, however, an enlarged therapeutic experience has learned to interpret. This is accomplished under the guidance of the free associations of the patient, which lead back to original experiences and impressions. Dreams also lead back to the same childhood content, whether they be the anxiety dreams which accompany the symptomatic disturbances of the psychoneurotic and reveal their meaning, or the pleasanter dreams of the normal adult. The ready enjoyment of a display of childish humor, certain vaguely perceived flashes of feeling which one traces to childhood days, the readiness of the adult to seek refuge at any and all times in infantile moods or outward condition, all of these testify to a close connection of the adult life with a childhood far richer in thought, feeling, and striving than can be accounted for in his clear conscious memory.

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* Bergson, "Creative Evolution," p. 5. "In its entirety, probably, it [the past] follows us at every instant; all that we have felt, thought and willed from our earliest infancy is there, leaning over the present which is about to join it, pressing against the portals of consciousness that would fain leave it outside. The cerebral mechanism is arranged just so as to drive back into the unconscious almost the whole of this past, and to admit beyond the threshold only that which can cast light on the present situation or further the action now being prepared—in short, only that which can give useful work."

They enable the sympathetic observer to gain an insight into countless half-concealed expressions of childhood's mental life, of which, to a large extent, the parent educator, or physician is only dimly aware. The adult is often unable to explain these sympathetically and bring them to the child's conscious acknowledgment. And because of the child's lack of understanding and the adult's inability to put himself in a state of helpful recognition, these half concealed expressions become only too often the source of a childish perversity or an indirect symptomatic anomaly.

Analysis of the child mind is essential. This analysis, however, cannot be one which studies a single mental element apart from the others. It must look back into childhood and summon to its assistance all that can be obtained from a study of the greater affective life that surrounds each individual history. This latter is inherited or is acquired from the individual's own ceaseless experience. It is the constantly active manifestation of the great force or energy of life which belong to the child no less than to the adult, which creates his sexual problems and conditions his striving with them. It must be sought for in the unconscious, as far as that can be penetrated, or in the constant influences arising from it which surround consciousness like a fringe, and are transformed by the power of intellectual control with active conscious progress in education and usefulness. If they are not thus transformed, they disturb this conscious progress by their more primitive egoistic character.

Nature never intended that our view of child sexuality should be as limited as cultural repression has made it. Morphologically provision is made during embryological life, not alone for that final function which is not matured until the period of puberty, but also for the more diverse manifestations which distinguish the formative years of early childhood. These early manifestations, from the genetic point of view, help to explain the diffuseness of the child's sexual interest. The closeness of the relation in the embryo of the intestinal, urinary and reproductive organs is a reminder of the closeness of function of these systems in the earlier forms which the human embryo recapitulates. Psychic experience is correlated with this morphological structure, if not to some extent in embryonic development, at any rate, in earlier evolutionary history. We can conceive it as imprinted upon the germ plasm constituting the inheritance of emotional reaction from the most rudimentary and elemental to the product of the slowly developed refinement and variety of sensation of modern human life. Traces of these earlier stages as well as a brief recapitulation of this course of development appear in the psychical life of the child. More specifically, it is to be noted that the intestinal, urinary and reproductive organs and their functions show a lack of distinctive-ness and separateness in the fantastic thought and in the practices of childhood.

The first appearance of the genital organs consists merely of an increase of the mesoderm beneath a thickened portion of the epithelium forming what is known as the genital ridge. Previous to this the Wolffian body with its excretory duct, or primitive kidney, has been formed from the thickened epithelial band. At about the same time a second duct, known as Müller's duct, is formed, which has also, at first, a nephridial function. This in the female becomes the oviduct while in the male it atrophies leaving only vestiges. The Wolffian body atrophies to a great extent; all but that part that goes to form the genital ridge develops into the ovary or testis. Vestiges of it remain in either sex to become suspensory ligaments. The enlargement and thickening of the walls of the passage into which the Müllerian ducts blend form the uterus and vagina. There is a change in the position of the ovary which causes it to descend to its adult position.

Early in development an evagination of the hind-gut forms the allantois, from which later the urogenital sinus makes its appearance. This becomes thickened in its walls; the epithelial tube lining it evaginates into the muscle to produce little sacs which thus form the prostate gland, while the urongenital sinus is now known as the prostatic urethra. Further transformations result in the appearance of the various features found in the adult testicles. The testicles or testes, like the ovary of the female, descend in the abdominal cavity, but while the ovary stops at the border of the pelvis, the testes pass normally out of the abdomen through the inguinal canal into the scrotum.

The cloaca is the common passage into which at first the intestine and the genito-urinary ducts discharge. At first, it is closed from the exterior by a thin membrane, but this, in the process of growth, is gradually broken down and absorbed, permitting an external opening to the cloaca through the anus. In front of the fossa in this cloacal membrane is formed an eminence known as the genital eminence which is soon surrounded by the genital ridges. On the under surface appears the genital groove continuous with the cloacal-anal depression. As this deepens it becomes bounded by the genital folds. These become the labia majora of the female and the scrotum of the male. The genital eminence forms at its top a rounded portion which later becomes either the glans clitoridis or the glans penis, while the rest of the eminence develops into the corpora cavernosa of the clitoris or penis and their surrounding tissues. From the genital folds are developed further the nymphæ with the bulbi vestibuli in the female, while in the male they become the intra-perineal portion of the corpus spongiosum, the intra-perineal urethra, and the enveloping skin.

Embryology teaches us thus how slowly differentiation takes place, whether in distinguishing sex features or in the separation of distinctive form and function. Nature apparently abhors the clear-cut distinctions which intellectual description has found necessary and strives to retain the unity of the continuous stream or flux of life. This standpoint of observation is therefore necessary in order fully to understand the fluid years of child life in its sexuality.

There is at first in the genital ridge no manifest difference in the male or female embryo. The determining factor which produces from this genital ridge the distinctive organs which characterize either sex is still unknown. Whether sex is established in the ovum or spermatozoon, whether it is determined at fertilization or during embryonic development, remains controversial.* There is, however, a period at about the seventh week when sex differences begin to manifest themselves. From this point on development proceeds normally to the complete genital morphology of the determined sex. The inherent bi-sexuality of the individual is evidenced, however, by the similarity in form and to a certain point in structural capacity of the male and female genital organs. Distinction of sex manifests itself further, however, in difference in the size and weight between the male and female infant, also in certain anatomical differences, which depend upon some influence exerted upon the growing form. These are supplemented by certain psychic characteristics which manifest themselves to some extent during the earlier years of childhood, but which receive their fullest development at the time of puberty. Growth and distribution of hair, growth of the female breasts, differences in voice and manner, as well as difference in tastes and pursuits are some of the indications of a development pursuing a distinct course in either sex.

Observation of the effects of castration upon male and female long ago established some interrelation between the genital organs and general somatic and psychic development. The study of the inner secretions of the genital and other glands, it is thought, will furnish a more precise explanation of this interrelation of the genital glands. These secretions are probably in the male the product of the interstitial cells in the testes and in the female of the cells between the Graafian follicles. These produce the genital hormones which act as the connecting links between the somatic tissues and the genital glands, exercising a direct and specific stimulus upon these somatic structures and thus increasing growth activity. They cause definite lines of development varying in the sexes, and they so affect the whole muscle and nerve metabolism as to produce profound and far-reaching alterations.†

On the other hand these so-called secondary sexual characteristics are probably not entirely dependent upon the genital system itself but are in part due to the hormones of other glands of internal secretion. Moreover the genitalia themselves seem to depend for their perfect functional development and activity upon the interaction of the whole endocrinous system. The relation, for example, of the thymus gland to genital development is fairly well established, the atrophy of the former being correlated with the growth and development of the latter. Likewise the thyroid, pituitary, suprarenals, and perhaps the pineal gland have their influence upon the development and maintenance of the genital organs in normal condition and functioning; while probably all the endocrinous organs have their influence through metabolic control, adapting the whole organism to its needs through the strictly genital function and through the secondary sexual characteristics. This is best known through the negative or deleterious influence exerted in a morbid condition of any one of these glands and the consequent disarrangement of this harmony. Then genital atrophy or hypertrophy may occur with abnormal increase or decrease of function and accompanying psychic disturbance or weakness.

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* Modern genetic studies show that sex differentiation starts with the chromosomes. (See Morgan, Wilson, and others.)

† See studies of Steinach and related biological experiments.

The knowledge that recent investigation of the glands of internal secretion has brought to light explains much of this abnormality of development, both hyperplasia and hyperfunction of marked character, and other disturbance of function as due to the increased or deficient action of one or more of the endocrine glands. Among these abnormalities may be mentioned aplasia or malformations of the genital glands, agenitalism, hypogenitalism in varying grades, or hypergenitalism with various anomalies of function of the genital glands. Since these are interrelated in the polyglandular system with the other organs of internal secretion, the syndromes include also those disturbances of growth and function which manifest themselves in other somatic spheres.

There may therefore be a condition of infantilism manifesting itself in a variety of ways,—small undeveloped form, sometimes graceful and finely built, sometimes remaining more distinctly infantile, at other times with an excess of feminine characteristics. Hypothyroidism, which results in myxedema or cretinism, displays frequent genital anomalies with infantile development of the genital organs. These are accompanied by widespread changes in the skin, bony tissue and other organs of the body, most evident in the thyroid, and effected through pronounced change in metabolism. Disease of the hypophysis results likewise in various disturbances of the genital system, causing increased development, degenerative or inhibitive defects, or dystrophic effects. These too are accompanied by marked somatic changes.

These manifestations do not belong strictly to childhood alone, yet they are of too much importance in the understanding of development and the interrelation of genital with bodily growth and normality to be left out of account. The broadest organic basis for the successful adaptation of the child to the demands of his sexuality can be established by bringing into the field, as active research is making possible, all these inferiorities of the endocrinous as of all other organs of the body and discovering the strength or the weakness of the physical equipment with which the child must express his psychosexual life. It is in part in such exact physical interpretation that definite cause can sometimes be found for the precocious appearance of menstruation, of erections, undue sexual activity, or of delay of puberty and maturation.

Too much reliance, however, placed upon a physical explanation of these anomalies cuts off the appreciation of the complete psychosexual background where lies the impulse engaged in its continuous effort to make its way to expression. In fact many things which are called anomalies in the older limited viewpoint find their place in this wider sphere as merely normal parts of development. They stand out as abnormalities only when they appear in more striking form, because medical and psychological insight has been so long blind to the reality of child sexuality. The structural basis of the abnormalities and disturbances which confront the physician in child life is sought first in the embryonic period. But these reach further than the more definite derangement of these special glands of inner secretion and their relation to genital development. The necessary view of the sexual life must be still broader.

Adler calls attention to the fact that any hereditary weakness which will become apparent in any other organ of the body must exist in the spermatozoa and ovule, and therefore that the place of development of both, and the whole sexual apparatus must bear its share in any form of inferiority. Clinical testimony bears out this relationship of genital anomalies in the case of diseases of the digestive apparatus, vascular system, or the kidneys, and also perhaps most conspicuously in the central nervous system. To these facts may be added the more direct influence upon the sexual development through the germ plasm, of the direct toxic action of disease in the parents, such as syphilis, or of alcoholic poisoning. Thus in a multitude of ways and in varying degrees, from the most pronounced diseased condition affecting growth and normal function to those subtle forms of disturbance which manifest themselves only indirectly and remotely, the child may enter the world handicapped in this important sphere.

Organic weakness, therefore, may form the basis of wide ranges of sexual anomalies. There may be actual physical stigmata or merely differences in childish behavior, which are included in sexual experience considered in the broadest sense. Disturbance manifests itself most extensively through activities and peculiarities which are to be found and studied in the psychic sphere. Here again Adler has expressed the matter when he states that the law of compensation is at work, not alone organically to build up a compensatory activity in a corresponding organ or in the diseased organ itself, in case of disease deficiency. Still more does the psyche itself strive for such compensation, mostly of course unconsciously. This attempt at compensation has, however, a twofold result. On the one hand it may be successful and attain to a greater strength psychically, and that means a greater capacity for constructive functioning through the organs which originally manifested the weakness; or it may occur in the progress of development in a related substituted form.

This is the path of sublimation. Upon it a lower function, through the original interest directed upon it because of pleasure attainment or satisfaction derived from it, may be made a source of effort and accomplishment upon a higher pathway which more completely serves the wider interest of cultural life. This higher pathway, however, must necessarily be somehow related through association with the former lower level. Masturbation for instance, may be cited as a single example. The child's early indulgence in this form of pleasure, influenced by early stimulated activity, and accentuated perhaps by psychic over-attention to the genital area, may by education and guidance be directed to certain forms of manual manipulation, such as kindergarten exercises, piano playing, typing, craftsman work, by which the tactile and motor activities earlier directed to unproductive pleasure find satisfaction now through constructive activity.

On the other hand the attempt at compensation may not be a completely successful one. Great concentration of attention upon the weaker organ or the functional deficiency, even though it be but slightly conscious, during the early years, or even permanently, may cause the pleasure seeking impulse to bring about an over-activity of the organ or function involved. Thus will the foundation be laid for sexual aggressions and perversities and, in the resulting struggle in the interests of culture, for the various forms of psychoneuroses which arise out of such conflict.

Certain obvious anomalies of childhood are frequently emphasized as signs of marked abnormality or evidences of a sexual precocity and are, therefore, considered among the exceptional phenomena. Centering attention upon these, however, has obscured the more important fact, that every child is in a sense sexually precocious, in that sexual matters are of far more concern to him than adults have been willing to admit.

Having therefore outlined the morphological and physiological groundwork of this period, in its normal state and in the deviations to which it is liable in the process of embryological and postnatal growth, it is necessary now to give more complete attention to the manifold characteristics of child sexuality, chiefly from the psychical side, showing the inter-dependence and widespread interaction of this phase with the entire bodily development.

Instead, therefore, of following the usual form of assertion that a child normally has no sex life, let us say that it knows as yet but little sexual differentiation. That is, it has as yet no defined notion of a sexual goal or a definite sexual impulse and function. On the contrary, it presents a multiplicity of partial impulses which have chiefly disappeared from the normal adult life because they have been synthesized and transformed into the completeness of the normal adult sexual aim.

The child with its abundant energy and plastic sensitiveness to impression comes into the world by no means sexually inert. It shares two fundamental instincts of the human race, or rather of all living forms. It must find nourishment for self-preservation, and it must reproduce itself. At first it neither grasps the complicated processes by which self-preservation brings to itself all manners and forms of nutritive elements, nor does it conceive of its power of procreation and its extended activity. It has the simplest form of hunger, and it realizes a need for self-expression, the assertion of its own inherent sense of power. Moreover its affective reaction is not alone the product of the sensitiveness of the newborn to a strange environment. As has already been seen it has within it a psychic response, developed through the countless ages of stimulus and receptivity, which belongs to its phylogenetic recapitulation.

Life is therefore already complex in experience and in reaction when the child arrives upon the scene of conscious activity. The best that can be done then in understanding the child's sexuality and aiding him in the control and use of it, is to begin at the beginning of his appearance among us. We must frankly discover what exists, and work with that. As Freud has said: "Indeed education remains properly within its assigned realm only if it strictly follows the path of the organic determinant and impresses it somewhat clearer and deeper."

The newborn infant has but few interests at first toward which to direct his attention. The world with its limitless environment cannot yet appeal to him through his senses as fully as his later activity will permit. Yet there is doubtless a certain intensity in the sensory appeal which he knows, which is born of repeated stimulus if not of the interest which his ego already makes its own. Hunger is present, and is very simply satisfied. Fulness of the bladder and the rectum occur and is readily relieved. Respiration has been physiologically demanded from the very start, vigorously too at first, and by a simple mechanism ready at hand the infant has found the means of appropriating the atmosphere about him to his wants.

Two important elements become thus a part of his limited routine. The first is the certain power which he has obtained over his environment. He needs air and takes it; he cries for food and obtains it. With the discharge of his excretions, which are a part of himself, he can impress himself upon his environment, altering that in a manner and drawing attention to himself through the act and its results. The second element is the contact sense in skin and mucous membrane, which has been relatively neglected in considering sensory impressions and resulting pleasure-pain. To this contact sense the infant has been exposed even during embryonic life, with how much distinctness cannot be directly stated. The contrast between the warm fluid medium in which he has lived and the cold dry atmosphere into which he is rudely hurled, awaken a violent protesting cry of pain. This same pleasant sense of the earlier pleasurable skin contact manifests itself frequently enough in child and in adult, if only in the unwillingness to leave the warm bed in the morning.

The baby is therefore already well sensitized to this tactile sense, the exercise of which he is scarcely limited in the early inactive months of life. It forms an important feature of all his daily life. It would seem that here too nature was provident of a stimulus which must play a large part, not only in general contact with surroundings, but in the specializing and intensifying of a form of sensation which must necessarily be one of the most important factors in the reproductive process. Thus even in infancy she lays, strongly and pleasurably, the foundations of sex life.

The warm bath repeats the intrauterine condition of environment. The taking of food, apart from the nourishment imbibed, inaugurates a new contact which is to be a source of endless pleasure. So at least we must judge, when we observe the continued suckling movements of the infant after the nipple is withdrawn, the quieting effect of the pacifier or of the voluntarily substituted thumb, or the phantasies of later years which express themselves in dreams, compulsive sucking habits, or gross perversions. The passing of excreta stimulates the mucous membrane. The urine, in particular, creates in its passage a sense of warmth and moisture both in contact with skin and mucous membrane, which is intensified by the moisture of surrounding clothing. This pleasure also is repeated frequently in dreams and phantasies, in the pathological habits of adults and in the enuresis of older children.

It is to be noted, in this consideration of the importance of the pleasure sensitiveness of the skin and mucous membrane, that the child is at that undifferentiated stage of development when he has scarcely begun to separate himself from his external environment. In the embryological state his world is very close to him, and there is no need for a sense of distinction from it. In the comparatively effortless existence of the early months of his life there are but few demands yet to develop a growing sense of separateness. His self and the not-self play but little part. The child then, even in his source of pleasure, has but the vaguest and remotest idea of another self than his own. Certainly he knows little of an external object toward which to direct himself. It might be said of him that he is pleasure seeking and comfort loving. In the wider sense in which these words are conceived, he may be described not so much erotic as auto-erotic. He is his own object toward which his efforts and impulses are directed. He himself is almost completely the source where his pleasure begins and the goal in which it ends.

Embryology and comparative anatomy, it has been noted, both teach the close association in development of the urinary and cloacal excretory systems with the reproductive. Infantile psychosexual history has much more to reveal in regard to the important influence of this association, as it continues in the early years, upon the child's psychic and social development. Here the connection is not so much one of growth as of association of function and of intellectual and emotional interest. Since emotion is the sum of previously experienced sensations, this emotional value may have a definite history in embryological and racial evolution, but that point must remain too obscure for present-day psychology to do more than admit it as a possible factor. Enough that the child is taught in his first experiences in cultural hygiene the importance of these excretory functions, an importance that is necessarily overstressed in the need to teach him in this fundamental way the first duties of care of the body. These activities, moreover, are such that even before specific education concerning them begins, the infant has discovered for himself the pleasure possibilities and the interest they may contain.

Auto-erotic as his pleasure sense is during his earliest years, it receives here a sum of definite stimuli which strengthens these zones of pleasure and preserves their importance in the activities of later life. This phenomenon is a part of the economy of nature, which works together with the discipline of the nursery, for the purpose of establishing the proper care and functioning of these excretory systems. Since the external reproductive area is so closely associated with those of urination and defecation, the sensory stimuli derived from the excretory acts, perform a twofold function. On the one hand, they assist to a certain extent in diverting attention and interest away from the genital organs proper until maturity has prepared them for function, while on the other hand they gradually direct attention thither and indirectly build up a pleasure sensibility.

This apparent paradox of development is one of nature's precautionary measures which is constantly in evidence in the psychic economy with which she works, and which often perplexes, unless account is taken of the ambivalence that runs through physiological and psychic activity. Antagonist and protagonist maintain the efficiency of muscular activity. Efferent and afferent impulses keep the nervous system in connection with the organism's environment. Tension succeeded by relief manifests itself in physiological action as well as in psychic states. In the same manner, it will be discovered, each phase of emotion has its corresponding phase ready to rise in alternate activity. Thus the child's attitude toward his world and himself is a constantly fluctuating one between two opposite members of a pair. Unless we take this into consideration much will remain obscure in sexual development and behavior and continue to defeat practical interpretation.

It is through such a balancing of opposites that function is performed and action takes place. Such a balance brings about a successful adaptation to life's demands and promotes health and normality. How easily this balance may be disturbed in the course of development, is shown in the psychical history of the areas under consideration. The over-importance of the anal region and the process of defecation appears in analysis of psychic disturbances to explain certain facts and conditions. Obstinate constipation begins often with the very small child, and manifests itself through a long life of neurotic history. Such a condition gives rise to the so-called scatological customs, without which normal function cannot be performed, which are rigidly adhered to with almost ceremonial reverence, and which consume much time and energy in a non-constructive fashion. These customs are traceable back to the pleasure obtained in infancy through the excretory function. They are usually discovered by searching in the dreams and habits of the neurotic, and confessedly lurk in the distress he entails upon himself. The actuality and importance of these things as sources of infantile pleasure and as means of self-expression, in lower form, receives support in the study of comparative development.

Anthropological study of peoples discovers many a confirmatory example of just such active interest in feces, the over-valuation of their miraculous power, and the attempt to control environment through their means. Procreative power is assigned to them, as to other excretions and secretions, in primitive and in infantile thought. Sympathetic observation of child behavior testifies to this interest. The infant just able to sit up and explore his immediate surroundings evinces active pleasure in feces. Culture has already imposed some repression on the older child. Nevertheless his interest in the fecal process is fully as evident in his shame, concealment, or his tendency to the secret laugh at various childish or accidental references to sound, sight, or odor recalling it. The small infant, eating its feces or smearing itself joyfully with them, neither of which is an unknown occurrence, is but following out the natural line of interested occupation with that which lies along the path of familiar experience and is associated with customary activity. This important principle borne in mind is the greatest aid to appreciative and helpful understanding and interpretation of the sexual development of these earliest childhood years.

Urinary interest takes an equal or perhaps even a superior place to defecation. The two processes in early childhood can scarcely be separated in functional activity or in the phantasies which grow up about them. These too find their counterpart in the history of racial feeling and thought, in the customs of the race as it passes through its childish stages. The racial mind, however, seems to have given the urinary function a somewhat advanced standing, as evidenced in its customs and beliefs, in its agricultural practices and in its mythology. Perhaps various factors which have built up the esthetic have been responsible for this; perhaps this process becomes more easily and thoroughly associated with reproduction as the race and the individual child advance in development.

The close association with the urinary function, both in activity and in the little understood vital process of the latter, is very evident in the childish sexual theories which are built up and often adhered to during adult life. The phantastic nature of these theories forms an important element in neurotic disturbances and is manifest in general psychic non-adaptability or more definitely in disharmony of the sexual life. It is manifest also in the frequency of childish transgressions and misdemeanors which, while largely occupied with urinary activity, have in them an obvious sexual element. Urinary contests are commonly indulged in during childhood by both sexes and certainly form a fruitful preliminary to later mutual or solitary masturbation. In addition to the sexual element, we have in this act a desire to express power and superiority and to influence in some way external surroundings. An example of the latter impulse, acting unconsciously, is shown in a certain neurotic patient who, while sitting in her car on the street, reacts to unavoidable delay by involuntary micturition. As an act expressing power and superiority, it has been used by both Swift and Rabelais. Gulliver extinguishes the fire in Lilliput in this infantile manner, while Gargantua repays the admiration of the Parisian crowd by drowning thousands and scattering others far and wide with the powerful flood of urine he sends upon them. The phantasy expressed with characteristic license by these two authors rests doubtless upon such features of general childhood experience, which if not expressed in acts, are present at least in inner phantasies.

Enuresis is a childish disturbance, annoying and distressing to parents, and impressing upon the child a sense of his disgrace and weakness. The unsocial character, in the widest sense, of such a disability, as well as the physical organic effect of its continued practice, compel us to take advantage of every possible means of understanding it fully. In order, therefore, to arrive at a complete understanding, one must look for the psychic as well as for the physical cause.

Adler has worked out his theory of organic inferiority with its attempted psychic compensation resulting in success or failure, particularly in this realm of urinary difficulty. He finds that with the inferiority of the urinary organs there is a corresponding weakness both of the central nervous system and of the sexual apparatus. So that there appear in the family history, not only all the diseases which directly affect the urinary organs themselves, but also functional and anatomical abnormalities in the sexual mechanism. These may be congenital anomalies showing themselves in the child's development, such as phimosis or adhesion of prepuce; or they may be functional disorders which have revealed themselves in the parents. At the same time there are reflex anomalies denoting the spinal weakness, such as spasm of the sphincter, which are often taken as the cause rather than merely as an indication of some more fundamental defect. Adler stresses particularly the evidence of segmental inferiority found in enuretics, which causes functional weakness of the urinary or sexual organs and which is connected with an inferiority of the lower portions of the spinal cord. This organic weakness, often a more or less obscure inferiority, inaugurates the process of psychic compensation which Adler has stressed as the attempt, at first unconscious, on the part of the organism to atone for its weakness and reassert itself. This may result in a psychic and organic strengthening, a building over of a weakness into some form of useful interest and activity which will advance the individual upon the path of development and social success. On the other hand this psychic attempt may fail, partly through the organic handicap and partly through the inability of the patient to turn this psychic attention away from himself and his inherent weakness. The attempt at compensation failing, there is a resultant increase in the dominance of the impulses toward pleasure and power which are seeking these organic pathways. At the best the method of the infant must be that of trial and error in the discovery of his environment, and in his experience and control over it. The organic weakness in urinary or genital organs but makes this more difficult and the results more uncertain.

As a rule the child's urinary experiments are perhaps not so obvious nor so conscious as those which have been cited. Cultural repression of these things in the course of training and observation make him, even at an early age, shy of revealing his interest even to himself. This would be also the conscious correction of the over-activity of the unconscious in its emphasis of attention upon the organs and functions concerned in the attempted compensatory process. Frequency of urinary dreams in the adult, an obsessive necessity for micturation at inopportune times, beside the often annoying insistence of children upon this function, all point back to infantile pleasure, to the exercise of his limited power and control of his environment. The earliest wetting of bed and clothing, the sensation of warmth and comfort, the demanding of the frequent attention of mother or nurse serve the infant as unforgettable sources of pleasure and satisfaction. They fade from consciousness under the influence of cultural repression and the crowding in of more useful interests in the course of education, but the pleasure thus created is conserved unconsciously, apparently in its original strength among the products of past experience.

Defecation and urination, therefore, are in themselves earliest sources of pleasurable sensation and activity. Indeed they continue as such throughout adult life, for they never entirely lose their share in association with the genital organs and function; while with but slight deviation from the normal, there is often associated with the sexual function a good deal of anal and urethral sensation. These functions doubtless bear a large part normally in the service of reproduction. Their over-valuation in infantile reminiscences, especially in psychoneurotic conditions, as well as their anatomical position, make it impossible to overlook their close association with the reproductive function. Their service lies doubtless in the centering of interest upon the genital areas, both by postponing pleasure activity until the individual has been physiologically and psychically prepared for the sexual act, and, gradually drawing attention to these regions and exercising them pleasurably, thus by helping to establish what Freud has called the "primacy of the genital zone."

These two important functional zones are by no means the exclusive centers of the child's early interests. Normally the whole body participates in the adult sexual act. It is not strange, therefore, that in the still more diffuse and undifferentiated dispersion of childish interest and sensation, in the lack of centralization, pleasure should also be discovered and resought in various other localities of the body. Definite regions are peculiarly fitted to be special erogenous areas, some by long association functionally with the sexual or other activities other by ease of access.

The mouth is closely associated with the anal and urinary regions as a center of childish interest. It is functionally active from the child's first appearance in the world. It is associated with love making and with many infantile fantastic conceptions of reproduction. The thumb also appeals universally as a most accessible area upon which to exercise the pleasure sense. Both the mouth and the thumb are closely associated with nursing, which, beyond the more strictly involuntary excretory functions, is, no doubt, the child's first pleasurable activity in regard to a local area. This is a nutritive function, which, in satisfying hunger, must give a certain sense of pleasure which increases with growth. A further enjoyment and satisfaction manifest themselves, however, in the delight with which the infant continues the suckling movements normally exercised upon the nipple within its own mouth or later with the thumb, when no nourishment is obtained. The flushed face and happy expression of gratification with which an infant withdraws from the suckling or falls asleep with the thumb in its mouth suggest the later appearance of gratification in actual masturbation or even, in a miniature form, adult sexual satisfaction. The same sort of manifestation can be observed in other children when they manipulate some other portion of the body, such as the lobe of the ear, for example, rhythmical rubbing of which produces the same pleasure giving, sleep producing effect.

It is easy, therefore, for the child's interest to be transferred from these earlier activities, from the fecal and urinary regions, and from tactile manipulation, to the genitals themselves. This change probably occurs in infants in less distinctive forms of activity. It is at the gradual awakening of interest in later years that it more definitely manifests itself. Masturbation can be accomplished in a number of ways, some of them growing out of the usual means of hygienic care, which in themselves arouse further interest in the genital organs. Certain pathological conditions also have the same effect, such as the presence of worms in the female child, irritation of secretions about the external genitals, adhesions of the glans penis or of the clitoris. Such conditions, however, are not always necessary to awaken the child's masturbatory activity. Various other methods may serve to arouse a certain pleasurable feeling in the genitals, most of which are normal and merely stimulate a desire even in extreme infancy to repeat the pleasurable process. Masturbation may consist in simply rubbing of the thighs together. The pleasure thus produced is of the same undifferentiated sort which is aroused by tactile stimulation of other parts of the body. This practice, however, like all diffuse sensation of this nature serves to prepare these parts, already specially provided with a sensitive nervous mechanism, for the later reproductive function with its more intensely erogenous character.

These earliest pleasurable experiences, important as they are in preparation for the later racial function, must, however, for a period, give place to other development. In other words, they must fall under *repression.* The interests of human cultural society demand that the fundamental racial function shall be set about with a vastly wider opportunity for creative activity, the energy source of which, nevertheless is to be found in the same instinctive realm of pleasure and power, whose basal expression, in its narrow sense, is the sexual. Therefore while the child's preparation must be one which will conserve and stimulate toward the mature sexual function, he must have the opportunity which either alternates with this function or in part accompanies it of training and developing many other portions of his nature.

With the acquisition of language and ability to enter more generally into human intercourse, it devolves upon the child to receive training both in intellect and in esthetic behavior. His auto-erotic activities must then of necessity fall into the background. At this point, the child arrives at that stage of his training, where any existing danger may be realized and where understanding of the continuity of development, with its manysidedness, may help to guide him on the right path and may come to his rescue when difficulty arises.

Society with its system of taboos has created certain aids to development. The child gradually falls under their influence, the result being that those things which were previously sought because they were pleasurable, are now put out of sight behind a rising sense of right and wrong, behind a feeling of shame, and a need for concealment. In many instances this sense of cultural restraint is so great that it will not permit a sufficient discharge, or "abreaction," of the repressed desire contained in the banished activity. Under such circumstances, an overcharged emotional experience keeps the repressed material alive for phantasy activity or future disturbance. It may seem presumptuous to believe that a small child would keep alive any considerable amount of repressed desire; yet analysis of neurotics of mature years has repeatedly revealed, without doubt, the existence of such infantile material, so active as to cause much of their later difficulties.

Close observation of child life in the earliest years would reveal much of the frank and undifferentiated interest which the child displays first towards its own body, and which is gradually extended to the bodies and the intimate life of those nearest to him, usually the parents. Following hard upon this period of unashamed frankness, the sympathetic watcher would recognize a period of more than adult restraint, characterized by a certain shyness and avoidance of the franker interest of the earlier days. This condition is exaggerated and therefore especially noticeable in certain neurotically disposed children. Others more normal in reaction, better able to leave past experience to the unconscious while turning healthfully to the call of external interests, become absorbed in the processes of education, companionship, the out-of-doors, and all the other various ways in which the external world summons the child to a new contact and experience with the things outside himself.

He is thus normally prepared for the next awakening of the sexual interest, which is perhaps stimulated anew largely by his externalized intellectual contacts. He is growing physically and psychically and is subject to a more defined and directed curiosity, not only in regard to his own body, and its origin and functions, but also to those about him, his parents, brothers, sisters, and playmates. There may be reasons directly due to his environment which arouse in him this curiosity and this renewed and more mature activity. Of course the advent of brothers and sisters stimulates question, though often through reticence, due to the influence of repression, this is but very vaguely and inadequately expressed. This reticence is too often made an excuse on the part of the parents for withholding that amount of accurate information on such processes of nature as his age and development would permit him to grasp easily. This instruction would save many of the phantasies which also form a part of the disturbing content of the psyche, and which interfere with a harmonious adaptation to the world and to the child's own power of development and function.

Evasions of this nature encourage the gratifying of curiosity outside of the home, where the truth is presented in distorted and repulsive form and where the child is subject to seductive influences from other children, and even from older persons. These outside influences are all too frequently the source of precocious sexual activity and often initiate the child into practices which destroy the normal course of his development and start him perhaps upon the course of some perversity.

For it is precisely during the period of childhood, when there is a great diversity of sexual manifestations that the foundation of later perversities is laid. To approach the matter from a different angle, it may be said that these perversities form a part of the sexual make-up of every child, and that they are merely the normal forms which the reproductive instinct takes in the immature human being. Every individual opens up and builds through his experience with the world these avenues of expression and surrounds them with the sensation and emotion which gives them value. They are truly perversities only when in the course of development any one or number of them form stopping places, when, so to speak , they become libido traps.

Libido is but a comprehensive name for the energy impulse so largely bound with pleasure, and therefore desire, which diffuses itself along the partial pathways in order to prepare them to take some part, as has been pointed out, in the final reproductive activity. In the course of development it is gradually withdrawn from the diffuse area over which it originally spreads, in order to be more completely concentrated upon the adult goal. Over-emphasis of any of these partial trends occurs only at the expense of such final concentration. Following any sexual maldevelopment, we find that there is not only an interference with the adult sexual function but also an inefficiency and inadequacy in the broader creative life of the individual. This failure to adapt oneself satisfactorily to life's demands is already manifest in the child where overgrowth of any one of these partial trends begins to reveal itself. This is hardly to be avoided in the process of development, which, with the child, it must be remembered, is a fluctuating process based like all human experience upon the principle of trial and error. It is important to keep in mind that reality, which comes both to the child and to the adult out of the immediate environment, is yet at the same time in the process of making, and that each individual has, more completely than has been recognized, his life to make or mar. Most true is this of the sexual life, which gives the key to the whole development, of which it is the source and foundation.

These disturbances of the developmental process do not appear alone. They do not occur predominantly in overt practices nor even in the satisfying of curiosity and interest in forbidden conversation with playmates. The most important part they play, as all anamneses show, whether conscious or occurring in dreams, consists in oppressing the child, either later or during the period of their exercise, with a sense of transgression and consequent separation from normal activity and thought. This interferes seriously with his adaptation to life, particularly the reproductive life and the finding of the love object.

What has proved itself more important than external transgressions and also more difficult to detect and to avoid or remedy, is the prolific activity of phantasy. The actuality of this phantasy life, its strange and protean content, and the reason for its hold upon the child, as well as its extensive influence down through the unconscious life of adult years, can only be understood if again ontogenetic recapitulation comes into view. Parents will admit with certain reservations that their children are little savages. If, however, the reservation is removed, it may be asserted that in a manner analogous to his embryological recapitulation, he has condensed in his childhood years, the childhood history of the race in its psychic aspects. He has rapidly to think, feel and act as savage and early civilized man have thought, felt and acted.

Mention has been made of the condensation of sensation in the psychic sphere belonging to the period represented by morphological intrauterine growth. This may be spoken of as the condensation into nine months of the history of millions of years. Similarly, in the period of conscious unfolding and conscious effort at adaptation, the child must hurry through the thousands of years which must be ascribed to the conscious childhood of the race. Therefore, the child has indeed to pass through the same animistic attempt to adjust the problems of life and to react with savage or primitive behavior. Add to this the fact, which probably exists, that he has a psychic inheritance which contains the affective vestiges of such an animistic method of thought and the effect which it has produced, and we realize something of the psychic material and the fruitful psychic soil to which the child attaches his own individual experience and upon which his own explanations are made.

Phantasies therefore are as real to the child as his overt acts and as the actual occurrences with which he has come into contact. This is due in large part to the form of thinking in which in his undeveloped years his mental life occupies itself. Like the savage he has not yet learned clearly to separate his own personality from the environment which surrounds him. His sense of self is ill-defined. Wish is as real as the hard facts of reality which make up his world. Since, however, his wishes are many of them incompatible with what that world demands of him or permits him, he must learn to fear it. Fear therefore becomes the conscious side of a desire which has suffered repression. Thus in this double form feeling manifests itself, and is still the main content of his thought life. Clear logical intellectual thought is a later acquired product. His attempts at reasoning, whether to make place for his desire or to repress it into the unconscious, enter into the phantastic and are carried out in terms and notions which are primitive and archaic. His intellectual activity is largely colored with feeling. It bases itself upon his experience. This experience, as has been pointed out, is, in his sexual life, concerned with many partial libido areas and tendencies upon which the phantasies multiply themselves. Externally also his behavior, his efforts to exercise his power and gratify his curiosity, occupy him thus. These are in part an expression of these phantasies; they are in part an attempt to discharge the interest along certain definite lines open to his limited experience.

The question of perhaps the greatest importance, even to the young child, soon comes to be the source of life and its manner of beginning. These processes of life are all about him. In his own home they first confront him in the natural question of his own origin, stimulated first perhaps by the addition of brother or sister to his immediate circle. The relation of his parents cannot escape him, at least as a general manifestation of sexual life. Too often also opportunity is thrust upon him to come into contact more intimately with the details of their life. For the limitation of psychological attention to the conscious world only has resulted in a neglect of childish impressions which has serious consequences in the sphere of the sexual development. It has been generally accepted and acted upon that the small child either does not observe or at most does not remember what it may have had opportunity to see and hear. We have forgotten the fringe of consciousness which surrounds any conscious act of attention. A little retrospection will convince any one that much more is perceived for example at a certain street corner than the actual object in the focus of attention. The rest, receiving no intensification, drops very quickly out of sight to be recalled perhaps with retrospective effort, or only to reappear unexpectedly from the unconscious under some other circumstances, which have brought it into the interest then at hand. Analysis of dreams has convinced us that infants too do not lose that which they have perceived even without the power of focusing intellectual attention upon it. It forms part of the increasing past which they are piling up and which influences their phantasies and activities of later years. Thus very young children do both observe and remember.

On this ground there is often actual material for phantasy and for those strange and inexplicable fears which form an obsession in child life, and which may persist through a later neurotic illness. Sense of smothering at night, terrifying dreams of attack, pavor nocturnus, for example, all, may be referable through dream analysis to the witnessing of the sexual act by the very small child. Since it is not understood except as a form of aggressive attack, it inspires an active fear and terror and yet is able to stimulate at the same time an underlying desire to share in experiences surrounded by so much mystery. Such desire explains in part the insatiable craving, particularly of the self-centered, neurotic child, to whom these things are more difficult of an external discharge, for a large amount of tenderness and attention from one or both parents. It is probably phantasy, occurring a little later in the child's life, that brings these remembered experiences of infancy into activating power.

There is, as an ingrained biological distinction, already a preference in the child for the parent of the opposite sex. This phenomenon is too universally confirmed by psycho-analysis as well as testified to by universally popular experience to be neglected as a factor in children's sexuality. It is but another evidence that the sexual life of the child is active throughout all its development even from the beginning, and places this preference for the parent in the complete scheme of the reproductive instinct with its division into two sexes and the attraction of these toward each other.

The actual influence of this early manifestation of sexuality upon child development is moreover a very important one. For remember already there is such a heterosexual relationship in the family. Therefore the child takes his or her part in it and asserts his or her individuality in it by actively turning to the parent of the opposite sex. Then he or she necessarily develops in defense of that individuality and its fundamental biological right, a sense of rivalry toward the parent of the same sex. The fundamentally biological is in fact often in opposition to the cultural. Therefore the child enters again into a struggle, sacrificing the wishes of the unrestrained ego to the one who is already in possession. The parent of the same sex is fundamentally a rival, as is the child who may later enter the family, particularly if the newcomer is of the same sex. If the new arrival is of the opposite sex, its advent often serves a constructive end, since it distracts the infantile emotion from fixation on a parent and directs it upon a substitute.

The history of human life has shown that childish impressions are so strong that conduct and emotional trends laid down through experience at that period tend to determine object choice, manner, and power of love in later life. All that goes to characterize and color the adult sexual life and its functional maturity are dependent upon the childhood years. The result therefore, of overemphasis and lack of harmonious development in the child's life is a fixation of libido upon some infantile trait and manner of gratification, with the consequent deviation from the normal. The infantile form of gratification being impossible of attainment in a cultural society, it too must enter the unconscious sphere and find phantasy satisfaction. This phantasy, however, assumes such proportions that it strives to find conscious expression. Relief, if it can be obtained must, because of social repression, be in a distorted form, which will not permit the true nature of the original impulse to be recognized.

Hence the suppressed wish expresses itself as fear. In dreams, the fear is symbolized by animals and robbers; in reality, by thunderstorms, snakes, and other terrors. The unconscious desire for the parent becomes in itself a fear of that same parent. Only thus can certain perplexing phenomena in childhood behaviour be explained, not only in the neurotic child, but also where development appears to progress normally. The child's reaction to discipline depends upon unconscious opposition to his own conscious inclination to accord with the parent's point of view. Neither he nor the parent understands why he stubbornly resists that which every consideration of expediency would seem to counsel. His unconscious antagonism against one parent, or his too ready attraction for the other, arouses in spite of himself a defense, which makes it difficult or impossible to receive discipline. More understanding of these conflicts within the child and their constant influence upon his effort at adjustment to life is necessary in order effectually to help him.

This state of affairs exists naturally in every child. Normally, however, it soon undergoes a wholesome adjustment under the influence of culture and social demands and the half consciously recognized advantages of a less egotistically centered life. The original impulses are transformed into a more useful workable form. In other words the reality principle makes its claim over the pleasure-pain principle which primarily governs the child. The child however, who is neurotically inclined, in whom this pleasure-pain principle has become over-accentuated, does not so easily escape. The bondage and the fixation occur more easily, so that the sexual development does not proceed harmoniously. Parents are to a large extent responsible for this condition. They have failed to understand the subtle influences emanating from the unconscious and by their attitude of ignorance have only helped to foster and strengthen them. They themselves have perhaps, from their own imperfect sexual and character development, made their children objects of the unsatisfied craving of their own inner egoistic natures. They have lavished upon the children the caresses and tenderness which have only served to strengthen the child's desire and to deepen the fixation. Or on the other hand, failing either to understand their own complexes of difficulty or those arising in the development of the children's sexual life, they have erred on the side of too great severity and have thus driven repression deeper and confined with it too much of the original childish impulse.

There is thus a broad background upon which the psychosexual life of the child is developing, out of which arise its rich possibilities, based upon the images and experience of the earliest important years. In it also lie dangers of too great attraction of its protective and love gratifying character, which when reality fails builds itself up through phantasy and forms a background for regressive desire at any period of difficulty and inactivity of later life.

It is necessary to examine further through what elements the sexual development proceeds upon such a normal background, and thus at the same time to find out where those dangerous points of fixation lie and how they maintain their hold against normal growth. We have said that the child's sexual activity and interest re-awaken when he has begun to take a more active intellectual interest in himself and his surroundings and to seek objects outside himself. The subject of sex differentiation stimulates his curiosity and his phantasy.

A tendency to peep and spy as well as to exhibit himself manifests itself in many ways. These tendencies must be recognized as a phase in normal development, although they may take any one of a number of courses in the child's psychosexual growth. They have, however, a direct service to perform first in the child's rightful acquisition of knowledge concerning his own body. Here there is opportunity for an open fairness on the part of the parent which will prevent perverted use of the child's natural curiosity, or the false modesty which is really a part of the repressive influence directed against transgression. In addition to constituting a phase of normal development, the looking and exhibiting tendency has a very definite function to perform in directing the child's knowledge and attention to his organs, in order that the "primacy of the genital zones," may be gradually established. Furthermore, the ambivalent tendency of looking-exhibiting must have its normal growth, since it not only forms an important accessory to the normal sexual activity, but also, through sublimation, extends the sexual to the greater creative life. The child himself later learns to peep into the secrets of nature, to discover the reality with which he has to deal, and to exhibit himself and his power in socially constructive work. On the other hand, the very strength of this impulse may, like other libido trends, meet with disaster through over-stimulation and over-activity on the pleasure and phantasy plane. Here again ignorance of the infant's pleasure responsiveness has fostered this tendency in its two phases. He is not only allowed to witness the secret functions of older people and other children, but he is made the object of familiar inspection and admiration himself. The impulse may be developed into a perversity, so that it occupies itself with forbidden exercise, and continues active on the infantile grade of sexuality. Or repressed into the unconscious by shame or other corrective reaction, it may exercise itself there in phantasy, missing sublimation, and recurring in neurotic fear, shyness or other symptom to disturb both adult sexual activity and social creative power.

Another ambivalence of impulse which belongs normally to sexual development comes into prominence in childhood. The aggressive tendency which belongs to the male, and to the necessary masculine element in the female, is easily subject in the child or in the adult to exaggeration and over-activity. It then becomes known as *sadism.* Normally, this impulse not only determines and guides actual sexual function, but it also enters into that wider creative activity which must be turned upon reality. Corresponding to it is its reverse aspect, which is known as the *masochistic*, which primarily represents the female attitude of submission and receptivity. It too is fundamental biologically to sexual function but must to a certain extent characterize the attitude of either sex. It, likewise, has its important place in the broader sphere of development and usefulness and belongs therefore to child life as a marked characteristic.

It is the fate then of either of these characteristic aspects to take its place in sexual development and fit both sexes for their respective procreative functions or for the special rôle they are to play in society. Yet either aspect may become of undue significance and may therefore form a factor in libido fixation and dissipation of libido to unproductive interests. This masochistic-sadistic impulse is most likely to manifest itself in ungoverned fashion in the uncontrolled years of childhood. Sadism is evident in the tendency to cruelty, teasing, aggressive insistence of the child upon his own way and to his own ends. It arises in part from the necessity of his making his way against a world difficult at best, which, if not actually aggressive towards the child, is often hostile in its lack of sympathy and understanding. He has not yet learned to regulate his own actions with an appreciation of the feelings and wishes of another or of the society in which he finds himself. This impulse is easily exaggerated in certain children to a marked cruelty, but this is doubtless due to an aggressive unconscious activity, in which reaction to phantasied wrongs, or actual injustice, misunderstanding and the like play a large part. Phantasy too in its turn plays upon the sadistic impulse building its fabrics upon it, often with a crudely erotic content.

It may thus be seen how closely sadism is allied in abnormal as well as in normal development with its counterpart, the masochistic tendency. That too is based upon the same phantasied injustice, is often an overcorrection of unconscious aggressiveness, and is found in the child who conceals his own aggression from himself and the world by diffidence, shyness, and sense of injury. And here too erotic phantasies run riot. His inner self extracts from these apparently negative reactions that sense of satisfaction which denotes the introverted nature, which directs all things toward its own ego. At the same time it satisfies its inner unconscious aggressiveness by an aggression upon itself. In this manner, these two phases of the same impulse play upon each other in the life of the child, forming either pitfalls in his development, or becoming useful avenues for the expression of himself.

These forms of childish sex activity pass easily into masturbation. This practice is probably manifest early in infancy where it is diffused through such tactile sensations such as have already been noted. At a later stage when there is a re-awakened interest in sex, attention is centered more directly on the child's own genitals and the sensations which can be produced there. Up to a certain point, masturbation may be regarded as performing a useful service in preparing the genital area for its primary function. Danger, however, lies close to such service, which, it may be stated, can be performed as easily and as gradually by the attention which is normally aroused by the hygienic cleanliness imposed upon the child, by his naturally aroused interest in sight and touch, and by the care with which he is taught to conceal and protect these organs. The temptation to go further and obtain pleasure by manipulation is a very real one. There is also frequently recourse to mutual masturbation, which closely allies itself to the looking-exhibiting impulse, and which seems also to contain something of the element of power expressed in emulation.

Much has been said about the dangers of masturbation. Great stress has been laid upon its injuriousness, both in the physical weakening of the body and the psychic injury brought about through its effect upon the nervous system. Account has not been taken in all this discussion of the injurious effects of masturbation of several things which cannot be rightfully neglected. In the first place, a more accurate knowledge of the nervous mechanism governing the sexual activity strips the whole process of much of its alarming mystery. The simple reflex mechanism involved is much less susceptible to injury than has been supposed, while the relative independence of the higher centers precludes the direct injurious effect supposedly resulting there.

Its real psychical influence can be truly appreciated only by taking into account that portion of the mental life where phantasies belong. In this sphere its results are deleterious, depending upon the degree in which they relate themselves to the course of development. Masturbation can in the first place, as we have seen, be looked upon as healthy. When, however, it becomes an end in itself and begins to absorb attention and activity, it is extending itself out beyond its healthy period and serviceableness. The child himself, particularly the older child, reveals this in his own attitude toward it. Of course this is due in part to the warnings and threats held out to him by older people or by the books from which he may try to get knowledge and advice. These set before him a frightful picture of physical and mental disability which result from indulgence in the practice. In the case of the younger child threats such even as that of castration or other form of punishment work direfully upon the phantasy life. Unconscious feelings of revenge, or it may be personal feelings of humility, inefficiency, self-punishment, actual castration phantasies, which in after years have a very serious effect both upon the normal sexual life and upon the whole character and personal ability, are the result.

Masturbatory activity shows its unconscious power even in the child life by its compulsive character. For with the best intentions and in face of threats and punishments it is still indulged in. This persistence in the practice denotes an inability to free oneself from the dominance of those phantasy wishes which have not sufficiently discharged their power before cultural restraint and cultural requirement caused them to be dropped out of sight. To this extent is the child unable to meet reality and make his adjustments to it. For this reason we find masturbation practiced by the neurotic, unsatisfied child, the solitary brooder, and the introverted child who shrinks from the hardships of environment. On the other hand, it may manifest itself in the otherwise normal child at periods of particular stress, at examination time or in face of a task of unusual difficulty. It goes hand in hand with a subjection to phantasy thinking rather than a healthy application to reality. It may, however, in the sensitive child be so early corrected that a substitute for it appears. This is not, however, the wholesome sublimation of the impulse into some occupation on the side of reality. It has still its hold upon phantasy thinking and though corrected in consciousness, it seems, nevertheless, to have a stronger hold in the unconscious. It manifests itself in some other more permitted form of auto-erotic form of gratification, such as toying with some other part of the body, fingering velvet, excessive fondling or love of being fondled, enjoyment of any tactile sensation in company with the brooding, imaginative life which marks off the masturbatory type from the eager healthy participants in the real, active world.

All these indications, however, supply the means by which this childish trend may be most rationally combated. The habit may be looked upon as a wrong distribution of energy, building up a form of auto-erotic gratification which, through the diverting of energy to the phantasy life, can never be sexually or socially productive and which exists thus apart from constructive activity. The important agency then is watchfulness, not lying in wait to frighten the offender by punishment and threats, but by seeking most earnestly to guide and redirect the energy to useful paths. Since this auto-erotic tendency is an early manifestation, which in fact normally precedes action upon external environment, there must be prophylactic care and direction so that the normal course of development may be encouraged away from a fixation.

The same principles apply to thumb sucking and other activities of the same sort, which are seen as substitutes for masturbation. They require tireless watching and devotion. It is necessary to employ the infant with toys or any attractive surrounding object so that his interest is absorbed and energy utilized upon the first objects of the external world of which he is capable of making even so elementary a use. As the child grows older the task becomes more fruitful. Employment for his active fingers, which may even be useful in itself, can be found in a variety of forms. His play, if subject to some guidance, may be sufficiently constructive, so as to make it a service to himself in the more serious later constructiveness toward which it leads.

Meanwhile the hold of the phantasy life may be definitely counteracted by the distraction of these external interests. There must be a profound understanding of the possibility of phantasy formation and phantasy activity of the child's inner life, mostly unconscious, which accompany masturbation. This necessitates a knowledge of those elements entering into the very young child's interests of which mention has been made. It is necessary to recognize how actively and prolifically problems of sexual relations of the parents, the infant's own relationship toward the parents, of desire on the one hand and of antagonism and rivalry on the other, forms objects of question and thought, soon crowded by cultural taboos into the unconscious. Urinary and fecal interest are also concerned, all forming material for confused interpretation of the relation with external environment and all infused with the emotional coloring which gives this phantasy life its vitality and its influence upon the outer life of the child or the later adult.

It is not enough that this is counteracted by the outer aspect of reality. One must realize that the vital force lies in the emotional life and be ready to release to a large extent the otherwise pent-up affect and so help the child free to exert his phantasy life in its true form upon external reality. This means greater frankness with oneself in accepting this actuality of such a life in the child, of these interests and the tremendous part they have played in earliest infancy and continue unconsciously to play. They can no longer be taken squeamishly and prudishly, terms which still hold their own, both in medicine and psychology. Those facts must be used which direct one's attitude toward the child in helping him to deal with them wherever they form part of his life. Over-emphasis should be avoided; the danger of that has been insisted upon in regard to the child's own experience. But each must be met as it represents a stage to be left behind in the child's education and growth, and the child must be guided not alone away from it but to appropriate its effective interest and appeal to a higher form of employment of its energy, that is, its sublimation. Thus, for example, excessive urinary interest, which might store up a pathological urinary activity, or harbor disturbing urinary phantasies in future development, is normally diverted into an interest in water and the various childish activities associated with it until, as has been suggested, perhaps an original over-compensation for urinary weakness, otherwise resulting in abnormal interest, may create the great navigator, or the keen analytical chemist, or the painter of water canvasses.

In the early development of the child there is a transition stage by which he passes from the early distinctly auto-erotic stage of an existence centered in himself as not yet differentiated from his environment. It is important in this study to bear in mind that every stage is a transition stage. The child reaches out through a distinction of himself from the external world to a finding of his objects of interest, libido, in that world. At first, however, as distinction arises he is still a self-worshiper. Only the self is at first very concrete. Now as differentiation and so also externalized interest have become more complete, his own body presents itself more distinctly as an object. It as a separate object is worthy of exploration, experimentation and admiration; it forms a discreet goal. Freud has likened this stage of development to the legendary tale of Narcissus who pined away with love of his own image, and has therefore named it the stage of narcissism. It is something of this interest, further than the purely autoerotic pleasure, which stimulates masturbatory activity as well as exhibitionism and even peeping.

Here also a transition stage is evident which leads on in development The chief identification, absorption into environment, in the child's history has been in its close relationship with the mother. This was only first altered by birth and the child's dependence was only then partially dissolved. Now as its body becomes objectified it has a strong feeling of identification with the mother to be overcome, or in the language of normal growth, to be outgrown. It is a sort of instinctive defense which leads the child along this course to fasten himself upon some further stage in the process which will form a safeguard against fixation upon the earlier stage, an aid to break away from it. The progress of his development demands that he reach this next stage and find a still more objective world. His next object choice is however conditioned by what has preceded. Therefore on this pathway he adopts first the person which possesses a body like his own. At the same time the mother identification and dependence upon her is a determining factor. He chooses such an object as she has loved. That is, he regards his own body through her conceived love for him and chooses thus an object of the same kind. This probably happens in the case of either sex, for with either the earliest relations with the mother are the same.

Thus arises the first manifestation of homosexual love. We have seen that in psychosexual as in physical development no individual is absolutely distinct as to sex. The wide range of the psychosexual sphere gives ample opportunity and manifests the need that this should be so for the complete social and sexual life. Both heterosexual and homosexual inclination belong in this complete life. Normally then the child finds his opportunity for freedom and development away from mother fixation with its dependence, or from the early love fixation upon the opposite parent of either sex, through a temporary escape to interest in its own sex.

Both this homosexual stage and the narcissistic, out of which it so inseparably arises, are of enormous social value. These too are subject to one of several fates in the progress of the child's history. Either one may prove a stage of too great attraction to the child, who from some external cause of difficulty combined with a disposition to turn inwardly to the self center, finds the regressive path easier than adaptation to reality with the sacrifice and change which that implies. Fixation therefore occurs upon either of these planes, producing in one case over homosexual inclination and practice or more frequently an unconscious homosexuality with its train of maladjustments and symptomatic disturbances. Narcissism may in its turn become an obvious ego characteristic or may also work unconsciously to retard the progress of the individual away from himself, or may project him into neurotic extremes of denial of self, depreciation of self, refinding of self in an extreme fanatical form. This has pushed the original attitude over into its opposite or over into an idealization which hides the true significance. None of these is efficient contact with reality. On the other hand, both have their important function in later sex life and in social usefulness and must be preserved and properly matured to develop into these forms of service. Narcissism realizes one's own worth and places it frankly before the sex object or before society. Human life, like all life, is essentially and fundamentally ego centered. It would be worth nothing and unable to develop were it not so. True valuation of the ego in the face of other egos is far different from the self centering of the introverted child who frequently hides these extreme forms of pathologic egoism behind a self depreciation and self effacement which serve to keep him, both as child and adult, from his share of the active work and responsibility of the world.

In love or in social activity there must be a normal valuation of self and realization of one's powers, even a certain aggressiveness, for the world does not wait for the uncertain character nor come offering its work or its rewards to such a one. The child must be helped to such an attitude of true self reliance. For this is his own escape from self centered and unsuccessful sex life and social life.

As to the stage of homosexual development and activity into which this narcissism naturally passes, it too holds its rightful place in the course of life's history. The sexuality of male and female, even in the limited sense of the word, is to a certain extent distinct. But even here this distinction becomes vague as it merges into the common characteristics of the sexual life and activity of either one. An appreciable amount of aggressiveness is necessary on the part of the female as well as the male and the various elements of sexuality, psychic as well as physiological, are fairly well paralleled in both.

In the social world this is still more the case. Here, especially in the modern world, both must be able to be assertive and capable of initiative, of impressing the self upon reality and reaching reality through the self. Passiveness and other feminine qualities which belong with it are necessary for both to display to a different degree, both in the sexual life and still more socially. Freud is, however, probably right when he makes the masculine quality of aggressiveness a primary possession of both sexes, called masculine purely, in this sense, because it is of such a nature and therefore the active creative element. "One might advance the opinion," he says, "that *the libido is regularly and lawfully of a masculine nature, whether in the man or in the woman; and if we consider its object, this may be either the man or the woman.*"

There is therefore ample ground for a homosexual tendency to manifest itself in development in natures so broadly and necessarily bisexual. It has its function in the mutual complementary attitude of the two sexes to each other in the strictly reproductive functions and in these as they are built out into social intercourse and service and completed by them. It is very essential for this reason that these bases should be well laid down as part of the psychosexual development of childhood. The more strictly sexual development is thus held in check in a certain degree up to puberty and beyond this, in order to divert the libido from the sexual aim until the older boy or girl too shall have acquired the many things which round out the sex life. This helps thus to build up the social intercourse and activities, the friendships and mutual coöperation within the sex, which are of inestimable value in a complex civilization. It also gives some outward phantasy discharge through a certain exercise of the love instinct where it will not yet be entangled in the more complex heterosexual relationships, which our civilization tends to defer to a later time and to more complete maturity.

The homosexual tendency, however, may be over-stressed, while forming an escape for the child already tending to fixation* on the inner regressive pathway. It is here only a partial escape. It merely shifts the same difficulty to another plane and leads into another sort of temptation on the path of fixation and regression. The child is early tempted here to mutual transgressions, masturbatory, forbidden conversations, obscenities, prying into secrets, which close association with one's own sex often makes possible, where it would perhaps be impossible to commit any similar trangressions with the opposite sex. The phantasy life comes to play a very large part especially where the nature is such that it is not easy to put the underlying attraction and devotion out into a more wholesome social activity. Particular watchfulness and understanding is needed here in the care of smaller children, their plays and conversations, often secretive. Lack of sympathy or even the slightest indication of harshness will only drive such activities under further concealment, or under repression, concealment even from the child's own consciousness, to work then serious disturbance in later life. It is the unconscious emphasis of homosexual tendencies and interests, rather than overt homosexuality with its practices, that works such subtle and far-reaching consequences. These things are fostered therefore by too great intimacy which allows of mutual confidences, physical embraces and love-making, very actual in many children. Here again is the danger of children's sleeping together and being thrown together in the intimacies of toilet and excretory functions. Every analysis is rife with experiences of this sort which have not only provided material for conscious and unconscious anxiety in after life, but have laid or strengthened the fixation in the love life upon one's own sex to the expense of the normal heterosexual development and therefore of full usefulness and happiness.

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* We use the terms fixation and regression to cover two phases of the same psychoneurotic process. Any one of these stages of growth in the psychosexual life may become a point of libido fixation for the reasons designated. As such they may confine a certain portion of the libido permanently, seriously interfering with the attainment of psychical maturity and freedom. Or they may be temporarily such libido stopping places, so that they have received an over-sensitiveness, so to speak, making them ready channels for the libido if later regression takes place. This is due to difficulty encountered later in life from which the individual shrinks into himself, reacting by libido regression to the earlier stages rather than by a healthful, adult grappling with reality. Both serve to account for psychoneurotic difficulties and probably neither occurs without at least some evidence of the other.

A word of warning may be spoken also against the danger of such intimacies and opportunities of contact in small children of the two sexes. The dangers which beset older children, in whom sexual interest has begun to manifest itself or in whom the approach of puberty lead one to suspect it, are more obvious and lead to more watchfulness. Although even in this respect parents and guardians are often singularly blind and timid about admitting or frankly meeting any such dangers. There is besides this a very real danger with very young children of misdemeanors committed actually in their "innocence." The fact of their intentions is a small matter, the after-effect through their phantasy life is very often serious and far-reaching. The small boy who compels the tiny girl sleeping in the same bed with him to arise from the bed, seat herself upon the chamber and allow him to urinate upon her, follows a childish caprice stimulated perhaps by an unconscious aggressiveness of his masculine sense of power. That such an occurrence came to light in later years as a dream association in the analysis of a neurosis reveals the power it had to the phantasy of at least the little girl, while the fact that she had carefully concealed the cause of her manifest distress at the time from her elders took its place in the secretiveness which bound her later libido and diverted it from success in her adult tasks. Wisdom and care need certainly to be exercised in regard to the close association of small children while no presumption can safely be made upon the native innocence and esthetic carefulness of the child himself, since he knows as yet no sufficient social restrictions of this kind. He is not only instinctively active to explore and discover all he can but to exercise just such normally aggressive tendencies. Close association, particularly sharing the same bed, gives abundant opportunities and throws children into such contact that erotic feelings arise toward each other prematurely and abnormally. Emphasis has already been laid moreover on the common practice of allowing the child in the bed of older people, particularly the parents. Love is active toward them in the unconscious in its erotic form, and this too receives undue stimulation and the child is driven more thoroughly to intense egoistic desire and the phantasies by which it satisfies itself. Besides opportunity is more easily given, although inadvertently, for the exercise of peeping and exhibitionism along the uncultural pathways.

It is necessary to gain a still clearer idea of the content of the phantasies which play their important part in child development. Their interplay both confuses and illumines the transition from one stage to another in this gradual psychosexual unfolding, but without proper enlightenment to the child they are carried over into his maturer years. There they are incompatible with intellectual knowledge of the processes and even with the normal adult emotions involved in the love life, and yet they live and disturb these because so colored with the emotional tone of childhood pleasure. In order to understand them and also the sexual activities that begin to come into play in the maturing child in connection with them, it is necessay to revert to what has been said of some of the infant's earliest interests and activities.

These the child engages in, in the course of his earliest education and later, when he is coming constantly into new experience of manifold sorts in this external world. He shares the universally human interest in life and death. The latter comes less to his notice, but the beginning of life is not so easily avoided. There is his own life to account for and that of brothers and sisters. Added to this and easily connected by him with this is the intimacy of his parents, specially sanctioned and permitted to them for some reason that he does not understand, but into which he inquires.

A most important factor in all this is the child's affective interest. These are not matters of purely intellectual concern. If so they might wait long for his attention. He is emotionally involved. They concern his happiness, his close relation particularly to the parents. His desire, his ego claim, is aroused though it is undefined. Being as yet an unreasoning ego he is sensitive, on the defense against infringement upon his phantasied rights, interference with his pleasure demands. Therefore there is both attraction and hostility, according to the way in which these wishes and his aroused interest are met.

This relationship is closest to the mother at first for both sexes, because of the complete dependence upon her through intra-uterine life and the early post-natal years. Yet it is commonly accepted that there is soon an inclination on the part of either sex toward the parent of the opposite sex. Examination of the inner life confirms and explains this. The fact of the existence of a sex life throughout the infant's development, in this broader sense of sexuality, gives it an element of attraction toward one sex and resistance to the other as a rival. Phantasy makes much more of this than conscious activity and thought do. The latter are soon subjected to the corrective influence of the cultural atmosphere, which puts restraint upon original ego impulses and also gives opportunity to turn them into a more healthful adjustable form.

Phantasy, however, carries on a separate existence to some extent doubtless in the most normal child's life. If it were not so the externalities of life would lose that value which comes from the richer emotional life in which phantasy has its activity. But normally these phantasies only form the background. They give tone and vital meaning to the intellectual development, which is the actual putting out into the real world of inner power and desire. The child to whom phantasy attains an abnormal value confines desire, introverting it to this inner life and seeks power as a consequence in the infantile forms already mentioned. Such power activity lies only in imagination, unreality as regards the external word, and therefore the child's life fails to comport with reality and its increasing demands upon him.

His own earliest activities, as we have traced them in their interest for him, are most closely bound with the persons nearest him. Both persons and activities therefore react upon him with an intermingled affective interest. One becomes involved in the other. As little is his early love separated from external factors as that of adult lovers. These factors are, however, for him as yet less distinctively divided into those openly permitted and those guarded by secrecy. This secrecy is nevertheless operative around him and he is very soon initiated into the process of repression. This does not however come about easily.

His curiosity is aroused and strengthened by the taboo placed upon certain things. He wishes to know more of those whom he loves best. He is forbidden certain things which have been most pleasant to him, and which had brought him into close contact with and dependence upon the loved object. This is the mother first of all, but with the female child this interest shifts to the father very early. This itself, as dreams reveal, soon manifests the character of a heterosexual attraction and doubtless it is often precipitated and fostered by the father's own disposition toward the small daughter or the mother's toward the son, in which either parent unconsciously indulges his or her sexual desire in the apparently harmless caressing and fondling of the small child, ignorant of the serious effect upon the child's inner life. Cases occur where it is the grandfather or uncle who thus becomes a father substitute and becomes the object of disturbing phantasies, or the nurse or some female relative takes the mother's place in the case of the male child. A female compulsive neurotic, producing sexual dreams of both father and grandfather, could trace an excessive skin eroticism, resulting also in skin disturbance, in part to a custom of this grandfather of sitting by the small child's bed telling stories and stroking and scratching the child's back.

The child is allowed free access to the parent's or other relative's bed in the morning, is allowed to witness the performance of the toilet, frequently with observance of the urinary function. This last is a frequently confessed source of phantasy disturbance manifesting itself in dreams, enormously exaggerated phallic phantasies and phobias in neurotic patients. One patient who had slept in her parents' room during her early childhood carried distressing actual memories of the sight of the father urinating, and had reacted with a repulsion to pears and all sorts of pendant objects, could not tolerate a hose in action, was tortured by dread of snakes, and had reacted by a half conscious recognition of the evil sense connected with these objects, against which she had elaborated a most complex and time consuming ritual of cleansing prayers. Of course this was only one part of an extensive system of complexes centering around the father, but that it was a very important part even the patient had recognized before any analysis was undergone. Such things reveal themselves as all too common to permit of the carelessness which prevails on the part of older people, first in presuming upon the lack of observation on the part of even the smallest child and the power of forgetting such occurrences. Experience, often of the most disastrous sort, strongly contradicts the basis of such presumptions. It should moreover arouse to a careful watchfulness and helpfulness in the education of the child in regard to these functions, which aid them to see the naturalness of them, but at the same time does not neglect the fact that for the child they are strongly emotionally toned as connected with his love and desire. Otherwise these go under repression undischarged and therefore still active. This must put the parent on his guard against exposures of the child to such situations and at the same time teach him to help the child more sympathetically and frankly through the emotional stress of such situations, when they sometimes accidentally arise in family life.

Experience also teaches that it is not superfluous to mention aggressiveness on the part of parents and other relatives toward the child's emotional life. Too frequently one hears of masturbation practiced upon the child by the father, the uncle, or even more perverted sexual activities. Hired care-takers cannot be chosen too carefully or children too well protected from opportunities for such manipulation on their part. There is however transgression unconsciously on the part of the careful parent or caretaker, which is due to ignorance of the polymorphous form of the child's pleasure activities, and is an indulgence of the adult's own auto-erotic tendencies, as has been previously said. The gratification of his tactile impulses works untold harm in stimulating and fostering the child's skin eroticism, making it doubly hard for him to release himself later from this masturbatory form of pleasure in favor of the adult gratification belonging to true functioning sexual activity. Moreover spanking likewise definitely stimulates the anal area, already concerned in pleasure activity and also in stubbornness and rebellion. These are likewise manifest in the very young child in his refusal and consequent inability normally to perform the function of defecation. It is a common matter of analytical experience to discover the stubborn, secretive, stingy and hoarding character bound up with rectal difficulties and anal eroticism and this is an expression of a stubborn hatred which appears at the same time in other character traits. When we hear so frequently the admission on the part of parents that they have punished their children in anger, we can more easily understand how the parent's own aroused sadistic feeling has produced in the child a similar reaction, in which pleasure and pain are intermingled in the way peculiar to the unconscious.

All these are of the phantasies which center themselves about the various functions and erogenous areas. Their proliferation through these agencies or through neglect and misunderstanding, as well as through the child's inherent disposition to avoid reality and continue in this realm, become actively destructive and disturbing factors for later life. They assume moreover definite active form about certain facts and questions. The questions of birth and sexual relations early become the center of these. The child's ideas are as vague and uninformed as those of the undeveloped savage who cannot scientifically connect the actual processes which occur. Like him the child has recourse to known experiences and builds out gradually from them and upon them.

It is not a mater of surprise or disbelief therefore that he believes babies can be created by anal processes. Jung reports the child who remained at the toilet in order to create a pair of horses for her wagon. The patient before referred to very definitely translated her first information concerning intercourse between the parents into an anal penetration. Fertilization by means of interchange of intestinal gases is met with as a childish interpretation and plays a large rôle in the wind symbolism in mythology as well as in medieval religious belief. The urinary function naturally becomes even more closely associated with the procreative function, and phantasies in which this takes the place of the latter are at the basis of phobias, for example, of accidental impregnation at the toilet used by male members of the household, an obsession of a patient already mentioned. This also acts as a disturbing phantasy which displaces actual adult knowledge and interferes with normal sexual function and gratification. It also causes much of the distress and inconvenience in the urinary function itself. Its involvement in children's enuresis has already been mentioned, while it is worthy of consideration in understanding adult genito-urinary diseases. Both this and the anal phantasies are probably largely involved in the sexual perversities manifesting themselves in the anal region or in regard to urinary function.

Another form of birth and reproduction phantasy manifests a peculiar frequency in child life. This is really more primitive because less directly connected with the sexual area. It is nevertheless an example of the hold which these earliest phantasies obtain upon the psychic life, in spite of later intellectual knowledge which to consciousness reveals their absurdity. The mouth was seen to be one of the first pleasure areas. As the area for the taking of nourishment, that is, of something which enters the body to produce some effect or change there, not unconnected also with anal interest, it attains a sort of significance which lends itself to reproductive phantasy. Therefore to children and savage alike, the procreation of babies is connected with nutritive processes and regions. It may occur directly by the taking of some food. Retranslated in the light of gradually accruing knowledge and the sense of mystery, children conceive a strange idea of fructification by the mouth. It has been confessed by more than one adult still inexperienced in sexual life, that they feared some mysterious influence through a kiss by which impregnation might occur. One such person voluntarily confessed to actual fellatio phantasies indulged in during childish masturbation, although apparently perfectly ignorant even at the time of the communication that such a practice existed. She had connected it, in characteristic infantile interpretation, with urination and felt, like so many a phantasy haunted individual, that she had been grossly guilty of a perverted state of mind peculiar to herself alone.

It is easy in the light of such voluntary confessions to appreciate Freud's use of the term "polymorphous perverse" for the sexuality of children. One may see in these characters only normal manifestations in the gradual growth into experience and knowledge on the part of the child. But there is an easy road of development into actual perversions, if development proceeds imperfectly into adulthood. With the average child, however, phantasies are often not so distinct, and therefore not so clearly distressing. Our enlightenment in regard to them comes by way of analysis of neurotics and their reminiscences, dreams and symptomatic substitutions in later life. It might be interesting to note in passing that the fellatio confession was made by a young woman suffering from a gastric neurosis for which there had been found no organic cause or therapy.

In general, we may say, these same phantasies occupy themselves more obscurely with the parents, their intimate life, the relation or unconsciously desired relation of child to the parent. They are thus probably active, though unconscious, to help us to account for many things. In the pronounced neurotic the phantasy desire takes a roundabout pathway, because of the repression due to the censoring on the part of consciousness. So in the average individual too it must reveal itself only in disguise and by a circuitous route. And in the light of these many and devious routes, who shall say which is the "normal" child? The term is after all only a pragmatic one, denoting the child or the adult who sufficiently breaks away from the power of phantasy to make a working adjustment to the more exact as well as the more useful things of reality. For this he abandons the circuitous pathways, or if in the imperfection both of human nature and external reality he finds a certain useful defence and aid in certain of these, he retains and uses them. When they trespass beyond the borders of usefulness they are abnormal and the child is thus far unsuccessful. Each must learn his own way and make his own adjustments through trial and error, not however without guidance and watchfulness.

These phantasies and their unconscious activity, created as they are out of desire, are, as we have said, primarily and fundamentally engaged upon the parents. They occupy themselves with wishes, love activities in these diverse and uncultural forms, in regard to the parent, usually that one of the opposite sex, and antagonistic ones toward the parent of the rival sex. They may be colored however with a homosexual tendency and therefore occupy themselves positively with the parent of the same sex. Sadger has reported at length a sleep-walking complex, with such an underlying homosexual fixation upon the mother. The patient sought in devious ways to repeat in sleep infantile peeping as well as comforting experiences with the mother.

In the more usual relation toward the parents, however, these phantasies are manifest in various ways. The resentment which the small child shows toward discipline is strengthened to the point of an unconquerable compulsion from the unconscious. We are all familiar with the violent antagonism shown by the little boy to his father's chastisement, or even to the father's imposed wishes, where no punishment enters in. We have seen the child struggle ineffectually with himself desirous of obeying, his intellect and reason accepting the father's better judgment while an inner rebellion takes possession of his action. Neither father nor son is conscious of the deep psychical resistance which makes them antagonists one of the other and defeats their conscious intentions. These are frequently more than the mere individual wish for independence and power, which are aroused in such situations of the child with either sex. These are aggravated in the case of the child before his father by the antagonism against his appropriation of the mother, his unique right to her sexually, intimately. He may also be influenced by a sense of cruelty imposed upon her in the sexual act, which the child has too often witnessed or of which he has otherwise gained some indistinct knowledge. In an unconscious way he is championing the mother as well as defending himself while resisting the father. The mother's discipline also fails for reasons not entirely dissimilar. Here the child has to utilize his rebellion and self-assertion to resist his own unconscious incestuous desires toward the mother, and the mother is likewise hindered by her own unconscious need of defense. Much of the difficulty and impossibility of discipline on the part of parents, or their later substitutes the teachers, would be simplified or obviated if these inner impulses were understood and acknowledged as the factors really at work. To follow them in their intricacy and distortion through the multiplicity and complexity of infantile phantasy, which translates them into terms of infantile pleasure areas and experiences, would as a rule be impossible. Yet so real and actual are these to children's inner thought and feeling, that consideration must be given to their existence, and to their actual content in the child mind. All this demands likewise of the adult that he stop to know and consider himself before he attempt the discipline of the child.

These desires, pressed down into the unconscious, struggling to find access again to the conscious life of activity, forbidden, causing untold conflict, are the occasion for yet far more pathological and unsocial manifestations. These have been recognized to have some sexual basis or at least a connection usually with undue childish sexual activity in one way or another. Their actual origin out of such a fundamental sexual foundation is however but barely suspected by many investigators.

It is only through investigation that one is able to appreciate the overwhelming force of these factors of infantile sexuality which have received such violent repression. The very resistance of psychology, and even of medicine, to the emphasis given to sexual origins reveals something of the strength of this repression. It concerns the most potent influence upon psychic and physical activity. Mankind throughout his history has been mostly occupied with the repression or the transformation of this tremendous force over into some other form of creative activity, some more widely useful energy discharge than the directly procreative one. Therefore in so far as the force has gone under repression in race or in individual it forms a mighty source of power. A certain normal repression allows this to be transmuted into sublimated activity and it thus serves the child for educational purposes, constructive activity in every cultural form. In the constitutionally inferior, in some form or other, where perhaps psychic struggle has compensated to the over-accentuation of some form of the sexual life or some erogenous area, repression must be stronger to atone or over-compensate for this. Earlier sexual transgressions, it may be only in phantasy thought, and desire, fall under more severe repression and yet their activity is not appeased. They are pathological and must break forth. Circumstances of environment, training, opportunity, as well as natural constitution, largely determine the manner. There may result only the milder neurotic disturbance already in evidence in childhood. Frequently, in particular environmental conditions or with particular constitutional tendencies, this assumes what are conveniently called criminal proportions.

Lying, particularly that apparently purposeless form which is known as pathological lying, may be recognized as truly compulsive symptoms, as may certain more violently criminal acts, all undertaken in contradiction to the individual's ordinary character and behavior. Herein they bear the common mark of the compulsions of the compulsive neurotic, for whom the compulsive acts bear a separate existence from the ordinary trend and choice of behavior. They are contrary to rational judgment and yet when they appear at shorter or longer intervals, have the individual and his better judgment in their power. They are an expression of the energy which for the time must be thrown over into the indirect and veiled, substituted accomplishment of the unconscious infantile purpose, determined and empowered by the infantile wish. In the compulsive neurotic there is still such complete social control that the symptomatic action chosen is usually concerned only with the individual himself in the manner, for example, of a private ceremonial.

He who lays his hand on the property of others, indulges in falsehoods which affect the character of other people, falsely accuses others to their hurt, or commits other offense against society, all in this apparently senseless, purposeless manner, must likewise be governed by an inner compulsion, which works more completely apart from judgment and control, and brings the dissociated wish into play. The unconscious tendency to pry into the affairs of father and mother, the sexual secrets, to lay his hand upon forbidden things, transforms itself into the socially more over-mastering compulsion to lay hands on other men's goods. Socially the latter may appear more destructive, yet we must admit that society, by the very openness with which it would speak of such misdemeanor, bears testimony to the greater taboo it puts upon the biologically more destructive transgression for which it stands, the entrance into sexual relations with the parent, the incest transgression. This has been a force against which the race has had to struggle throughout its thousands of years of history, until under the force of this long repression its compulsive power to determine substitutive acts may be to some extent understood. Lying in the same way is an evasion of true knowledge of one's own deepest wishes so culturally and biologically forbidden, yet so affectively strong as they lie in the innate racial and individual tendency to sink back to the source of things. For this lies at the basis of the incest struggle throughout all forms of life. This is strengthened in each human individual by infant dependence and the opportunity given for the exercise of the innate tendency in so many directions, through phantasy, or through the early experiences.

Ordinary sexual transgressions of children are not the crucial point of this compulsion. It is a fact that they are not so difficult to discover. They even form part of the content of pathological lying at times. It is the deeper infantile content which, because it is concealed from the individual himself, works such power in his life. One of the discoveries of psycho-analysis most significant for therapeutic success through readjustment is the displacement of affect. The original unbearable impulse is so utterly inadmissible to conscious thought that even the anxiety and disgust occasioned by its unconscious existence, which might seem to make amends for it, do not do so, but must be transferred over to some more accepted permissible activity or event. How much more so is the desire itself in the form of a compulsion, such as drives the child to such manifestations as pathological lying or stealing or even sexual transgression! Even this last, though not entirely permissible, may act as a cover for deeper activities which lie much further from conscious thought and understanding, in that realm of the unconscious which is denied by most thinkers. Entrance into this deeper unconscious comes through investigation of the emotional life as it was built up in the earliest period of childhood and through consideration of the earliest impulses and manifestations of the race. For here these deeper forces, the incest impulses, have had to hide themselves deeply and securely and therefore it is that the desire and the reactions following have had to be carried over upon something else. The force of the compulsion lying within is due to the intensity of the pleasure to the individual not so much in the direct incest wish, which only in rare instances manifests itself to the child in its concrete form. Instead the child knows it in the many elements which go to make up infantile sexual interest and activity. Spying upon urinary and anal functions, curiosity and peeping into all the intimacies of the parents uncluding the sexual act, archaic birth phantasies, all these form the subject of protean phantasy as well as of minor transgressions. They manifest a deeper desire at bottom, the fundamental incest complex, and because concerned with it are of deepest significance and must go under the greatest repression. As a result they form an overwhelming compulsion which manifests itself in the nature and insistence of the overt "criminal" acts, so inexplicable from the rational point of view.

This compulsive quality arising out of a repressed sexuality would probably be explanatory of much of the crime which is committed. Crime as we all know manifests itself in an enormity of sexual transgressions and in all its forms is usually more or less plainly associated with the sexual life. It is probably even to a greater extent and in its compulsive character the expression of such a primitive and infantile form of sexuality that it is not permitted the direct expression of itself even in actual sexual form, but the impulsion within it must manifest itself some other way.

At the period of puberty, when phantasy life is more abundant and when friendships and adorations among one's own sex have a particular function to subserve, as has been mentioned, the child's inner life must be watched sympathetically. Phantasy here is especially active and may become a source of danger, also strengthening an earlier difficulty in getting out to normal contact with the world. It builds up instead a baseless and unserviceable fabric of daydreams, false pleasure rather than reality. For both this danger and the same sort as it appears also in the earlier years, school activity, interest in intellectual pursuits and social pleasures in connection with the objects of friendship provide healthful sublimation. Work and play, the latter especially in the form of wholesome athletic pursuits, receive a special incentive from the forces at work and form as well safe outlets for these same forces. Normally, as has been reiterated, these various tendencies enumerated belong in the gradual centralization of the child's psycho- sexual life upon the normal sexual aim. It has been seen how impossible this is of separation from his general development into the breadth of an intellectual and social life. Yet the growth of the sexual impulse is also a thing which can be distinctly traced as a particular phase of such development. The tendency of medicine in the past has been to treat as pathological, and evidence perhaps of perversity, any precocious physical or psychical signs of sexual maturity. Genetically, we see how impossible it is to expect any hard and fast adherence to fixed rules on the part of nature.

Climate, custom, general bodily vigor and many other factors exercise their influence on various races and peoples as to the age of maturity, and probably upon individuals likewise. More emphasis has, however, been laid upon them, for instance in the matter of appearance of menstruation in the different races, than careful research seems to warrant. Individually at least it can be believed that many factors enter in, which are largely psychical. The close association of menstrual irregularities, and also of the abnormal activity of the male genitals, with neurotic difficulties convince us that there is a very close relation also in normal activity between the first appearance of these functions and individual psychic life. Even normally, phantasies play their part also as does likewise the particular direction of psychic interest. Phantasies must occupy to some extent every child mind, and their content is an indication of the special libido inclination. Health and normal development depend on the ability not to become entangled in these but to keep and develop such a free and growing contact with reality that adjustment follows naturally, and the pathway is opened for the phantasies to escape into the corrective influence of real thinking and real activity.

The approach of puberty normally is marked by the appearance in the child of the characteristics of adult sexuality. Physically there are marked external changes in both male and female. In the former the change of voice is noticeable from its childish character to the deeper tones of the adult. In both sexes the genital hair makes its appearance with the first indications of a beard in the male. The breasts of the female begin to enlarge and there are certain other indications of the rounding out of the whole form to its adult completeness. The secondary sexual characteristics become more pronounced as regards either sex both in the physical sphere and in the psychical. Changes in the latter show themselves in differences in tastes and inclinations and tendencies which more distinctly now separate boys and girls from each other.

The most significant and comprehensive change that occurs, that which includes all the others, and in which they participate, is that of the change of the sexual aim. This means that the elements of the infantile life are now normally directed toward the adult sexual aim and gathered into the one main impulse. The primacy of the genital zone is now established. The indications that the normal functioning is now prepared are the appearance of menstruation in the girl and the erection and discharge of seminal fluid in the boy. The maturation of these functions is, it must be remembered, a gradual process and there is great variability in their manifestation. For even wider than the range that exists for differences in this physical maturation is the limit within which psychical differences may find themselves.

There has been a tendency also here to look upon the phenomena of the early appearance of erection or other phenomena of sexual activity as signs of a precocity which was abnormal or pathological.

A manifestly concrete sexual interest has been regarded in the same way. There has been a strange disregard of the universality of such interest on the part of all children, though with some it has become more evident than with others. It cannot be denied that excessive indulgence in masturbation, or other sexual practices which stimulate activity and conduce to phantasy formation, influence this early manifestation of sexual maturity. These may be due to seduction on the part of other children or older people. They are also the effect of phantasies aroused in the child under the influence of erotic reading, erotic pictures, particularly perhaps the stimulation afforded to phantasy by the moving picture theater. The influence of these upon the sexual life of the child is perhaps greater than can be appreciated unless one realizes how the sexual life extends into every portion of child development, and also how every part of the child's make-up and experience is permeated by the sexual and under the control of the broad sexual impulse. The moving picture has therefore a useful purpose to serve when it presents in the simpler form comprehensible to childhood the efforts of the individual and his striving with hidden complexes in a manner which is helpful to all who as spectators unconsciously participate. For the true artist is he who touches thus even in a simple story or play such fundamental problems and then connects them with reality by working out an artistic solution. This makes it, by just so much, easier to take up these problems and solve them in actual life. Such is the mission of the adult theater and such may be that of the miniature form accessible to the child. Aside from this of course is the more strictly educational service which a special form of pictures presents.

Few moving pictures however fulfill this ideal. In the first place they administer a too constant excitement and stimulus to the child's already active phantasy life. This is because of their easy accessibility and can only be counteracted by a more reasonable, careful regulation on the part of those who have the child's activity and attendance in control. The constant stream of excitement, lacking for the most part also the touch of truth and application to reality, makes actual conditions seem unreal and undesirable and therefore renders still more difficult and unattractive the adaptation to reality, which alone grants success to the child and makes his sexual life a healthy part of such adaptation. Besides, the material offered by the screen has much over-exaggerated covert sexual reference. Often this is in certain infantile traits such as the child himself has often difficulty enough to transform from the gross form in which he has first known them and in which they still press upon his interest and attention preventing their sublimation.

Immoderate excitement and over-stimulation tend also to force the child's normal development and thus to sidetrack the energy which would otherwise be directed toward unification in the adult primary goal. The genital life which is now approaching maturity and the cultural life which proceeds apace with it must be guarded and guided even while it is left free to pursue its natural course. An early understanding and sympathetic recognition of the natural quality of all the earlier and later sexual manifestations is the best means of establishing and maintaining that confidence in the child which will lead him to seek and obtain help not only, but will keep him from much of the danger of phantasy reaction which follows the thwarting of perfectly natural impulses and manifestations. This is the attitude essential for parents and for physicians, from whom sympathetic aid should be obtained.

Both boys and girls should be thoroughly prepared and in sufficient time for the marked phenomena which will occur at puberty or often at an earlier period than would normally be expected. Too frequently we hear of the fright which these have produced in the unprepared child, and aid has been sought in companions, equally unprepared for skilled instruction, in spurious literature or from unprincipled sources. The interest and attention aroused through fear and ignorance have led to pernicious masturbatory or unhygienic practices on the part of the individual alone. It becomes much easier to instruct or to aid parents in the instruction of their children and their initiation into the functions and self-knowledge which belong to their coming adult development, if the genetic point of view of their sexual development is the one attitude of the parent, instructor or physician and has been held practically before the child as well.

The maturing of the normal functions may then be seen to be possible at varying periods depending on many factors in the child's life. Sexual precocity itself becomes a less formidable term for there are numerous elements which can be taken into account to explain and understand the undue manifestation of sex activity in childhood. And this means a more rational control and prevention of such undue activity through the wider psychical approach. An erection occurring at even a very early age may thus find ample explanation in the overactivity of the child's phantasy, perhaps depending on some special psychical relationship. Such a phenomenon has been looked upon as either a sign of a childish perversity and depravity or attempt has been made to explain away its sexual significance on the ground that there is no sexuality in the small child. Love phantasies have been thought to be utterly foreign to the normal child and if present to constitute a symptom of something abnormal and perverse. The attitute however which sees no break in the continuity of the love life from the beginning of responsive emotional life on through the years of maturity, is not surprised nor discomfitted at the appearance of its phenomena in the years of child development. It also recognizes in the dynamic and genetic conception of the love life no distinction in its character, merely a difference in form of its application to the different love objects upon which it successively fixes itself. As to its intensity, there is reason to question whether in the impressionable and necessarily experimental years of early childhood it does not outdistance that of adult love in a way that colors and influences all subsequent success and failure in love.

If it is so aroused in the child, it becomes understandable how variable may be even the activity of the genitals and their period of maturity. Some slight genital weakness, organic or acquired or accentuated through masturbation, may cause an earlier response of a physical nature. Or this may occur under some particular emotional stress. Erection in fact frequently occurs for the first time or repeatedly under the strain of an examination or of a difficult task, sometimes with accompanying pollution. Or it has been known to manifest itself in a still smaller child upon close contact with the mother when she assisted the child in its bath. It is not necessary or wise perhaps to explain to so small a child the significance of the phenomenon but it is important that the mother should know something of the power of a love which is sufficient for so marked a stimulus. She must guide the child away from an early fixation, which would tend both to a prematurity, through phantasy activity, and to further introversion to phantasy activity because of the impossibility and therefore unreality of such a sexual object. There is with the girl analogous clitoris activity, which is stimulated in the same way by phantasy or by actual manipulation through masturbation or through forms of seduction.

When puberty is sufficiently established to permit of the appearance of the orgasm the child should already have been instructed as to the significance of this and concerning his functional capacity. As much as possible his activities should be turned outwardly through his studies and companionships and sports so that he may develop his general cultural abilities and utilize the new accession of force and energy toward an active constructive life. For this should normally be the trend of his physiological and psychical development and impulse at this time. Otherwise this energy, seized upon by fear, shame or other phantasy activity, which becomes phantasy lure, finds a refuge for itself in an introverted life, fails in its adjustment to reality and this critical period becomes a turning point in the wrong direction. Neither the sexual life in its stricter sense nor its broader influence upon character and culture finds the right road out. The boy should understand the naturalness of the awakening of erotic emotions hand in hand with his new ability to love actively, but at the same time his further duty of sublimation that his love life may conform to social requirements and cultural necessity, which gives it the greater breadth. He should be made to realize that the newly awakened force in him is only an advanced expression, now in matured form, of that same creative power which has always been active in his life, and which has a function to perform of the greatest significance. The racial meaning of procreation should be brought to his attention no less than the broader social purpose which is supported by it, but which in a cultural society exceeds it. He should understand clearly the nature of the sexual impulse, its mode of activity, the value of the dream as a means of discharge of the natural impulse and the consequent nocturnal pollution, which is too often a source of fear and distress and leads through them to disaster. For fear and anxiety again center the attention upon the unconscious phantasies or the conscious erotic tendencies and increase their hold upon the imagination. This leads in turn to masturbation or other practices or it may be to premature heterosexual activity, which only further aggravates the trouble in a vicious circle. Too frequent phantasies, manifesting themselves in too frequent pollutions, are best met and controlled by a sympathetic recognition of this phantasy life and the enormous part it has played in the years preparatory to puberty as well as in puberty itself. Sometimes it is necessary to submit such unconscious phantasy life, manifested through neurotic disturbance, excessive anxiety, inability to concentrate upon school work, to an actual analysis, when unconscious difficulties come to light and are discharged, which were seriously interfering with sexual and cultural adaptation.

The same also holds good with girls. Although their external manifestations differ to some extent from those of boys, they too pass through similar anxieties and disturbances. They are also left in ignorance to meet the function of menstruation and react in much the same way. The sexual differences become more apparent at this period, so that the element of retirement and passivity are in evidence at this time. This makes it more difficult perhaps for the girl to escape the retreat into phantasy life. Here she finds refuge from the new sexual activity, which, though she is partly unconscious of it, takes possession of her too. She fears her erotic impulses and at the same time they have a fresh power over her and attraction for her. Therefore she carries them largely into phantasy.

Besides the establishment of the functions of ovulation and menstruation necessitate some physical readjustment, but more upon the psychical side. In both nature has been preparing the way, yet for both some special provision must be made. Too much stress has however been laid upon the physical side so that it has been presented to most girls as almost or quite a pathological function and they have responded to it accordingly. A moderate amount of rest and care must be exercised, particularly at the beginning, until the organism is accustomed to this new function and the metabolic changes it necessitates. Yet it is a normal function and concerned with a special creative activity. Therefore it should be looked upon as such and as participating in all the activity and incentive to activity of which healthy woman is capable. While it is connected with a special function and as such makes a special demand upon personal hygiene and personal attention, it should not be brought to this attention in such a manner that it becomes an end and aim of phantasy life also.

The many disturbances of this function which appear from its onset or in early maturity and continue perhaps throughout the reproductive period, necessitate a serious consideration of the parent's and physician's attitude toward this. We do not need to study the history of menstruation and the part it has played in the religious and superstitious thought of the world to realize that it has a tremendous emotional significance yet the customs and beliefs attaching to it throughout such history throw a light upon the extensive psychic value it has to the unconscious and the part it plays in the emotional aspect of the reproductive life. It becomes for the girl or woman the outward sign of her reproductive power, as it accompanies her period of physiological readiness and ability to reproduce. It is therefore an expression of the fundamental wish which controls a woman's life. Any injury to that sexual life therefore, in an imperfect adaptation, failure to realize its fulfilment, missing of the love object, sexual dissatisfaction, readily manifests itself in disturbance of this function. Dysmenorrhea, amenorrhea, many of the disturbances difficult of explanation and treatment by the physician or surgeon are often amenable to the analytic interpretation and readjustment of the psychical life and the clearing out of the phantasy life which is responsible for an unconscious psychic misuse of this otherwise normal function. This too should be given full consideration in the understanding of girlhood at the time of its entrance into puberty and forms another reason for understanding as completely as possible the phantasy activity which disturbs the development and beginning as well as continuance of function. Here too rests the necessity for direction of the child's interest and activity upon external things. Not only thus is the libido prevented from phantasy fixation under the special stimulus of this recurring function, but the function itself and the entire reproductive process of which it is a part is brought into healthy normal relationship with the whole of life.

This is in reality the aim of all work in child education, the goal of understanding and the end of medical treatment and readjustment of disorders in the sexual development of the child. This is the reason in fact for a broader and more genetic attitude toward the sexual life, acknowledging its reality and recognizing its extended influence through the whole of the child's development. It is to enable the child to take his part in the reproductive life of the race and even more in our complex civilization, in the constructive work of society. This depends upon the knowledge of his own forces and the use of them, guarded and trained to adult functioning in the strictly sexual sphere and infused with the primal power of the sex life and instinct in the wider social sphere.

This means the elimination of these infantile elements and activities as means of dissipating the energy and creating a world of unreality instead of the real world of action and conquest. It necessitates from the earliest infancy to maturity watchfulness of these traits, with appreciation of the pitfall that each may present unless the energy, which is rightfully occupied at each level of development, receives sufficient opportunity and incentive to continue its course to the next stage. Such continuous progress can only come about if each infantile manifestation is reckoned as a thing of value in itself, as a transition to something further, and yet remaining to contribute a subordinate part to the transformed libido direction of puberty. From the earliest moment then the real world must be made the object. Every one of the bodily functions, each erogenous area, signifies just so much libido power behind it and just so much opportunity for this libido to come into contact with the possibilities for constructive activity which lie in the environment. The thumb finds its external usefulness first through the simplest toy, and is saved from an auto-erotic, phantasy building service. The child learns that instead of maintaining a useless tyranny over other individuals through his necessary functions, and therefore keeping himself a slave to unproductive and primitive exercise, they are only a part of the hygienic care of his body. This body is a serviceable tool, his only means of actual power, which is by just so much unrealized as he allows its pleasure seeking to have the mastery. Thus by watchfulness and direction the tongue is trained to language, the hands to the production of something which alter reality, working upon it constructively rather than finding an egoistic pleasure upon the self. The curiosity, which must naturally find its first stimulus within one's own body and then in the immediate family surrounding, is also accepted as natural and quite within the child's right. Only in this way can its value be secured for further direction at once out upon the external world. For always behind the external interest works the actuality of the power which has its source within the individual. The ego center can never cease to exist as of prime importance, but it is saved from itself and given opportunity of power by being gradually but continually thrust out from itself.

Sadism and masochism rightfully controlled and early directed are the child's lawful and useful weapons of attack and defence in the adjustment necessary to life. Here again the inherent value of these forms of self-expression must be admitted in order to help the child redirect these from his original cruelty, teasing or introvertive self-wounding and self-depreciation. Without peeping and exhibitionism he will not discover the hidden sources of energy and opportunity in the world about him. Nor will he know how to insert himself into this world in a truly expressive and impressive fashion. He will rather be overcome by the original infantile form of the desire to spy or to be seen, which because of its asocial character will set up useless and preventive defences.

The parent complex need not become a disturbing and pathological one if also reality is made a sufficient corrective here. It is the very foundation of psychical economy that the child shall emerge from so intense a miniature emotional world into the larger one. He must of necessity retain a close bond with the affective side of nature. He must also hold before himself, because he is an affective creature, a group of ideals personally embodied. Yet it is most essential that there shall be no fixation upon them but that they shall be in the course of racial history only spurs for his own endeavor, models for emulation, but more than that stimuli for independence and advance in his own particular course. Here is the greatest demand upon the wisdom and the adaptability of the child's guardians and directors, to step aside and give him this opportunity.

Just here too medicine, dealing with the perversities and anomalies which appear in the course of his development, must consider them as only disturbances and disabilities in the effort to get himself into his real adaptation with the world. They must be genetically interpretated and then their place as merely misapplications of an energy seeking to come to its own will be sympathetically met and aid be given to a free adjustment.

In the same manner the child's friendships, his homosexual development as well as his early inclinations to the other sex, come in for understanding and guidance. Undue secrecy, childish obscenity, masturbatory practices or even more serious misdemeanors will be met in a spirit not of that severity and condemnation which produce reticence, repression which makes for compulsive even criminal symptoms, shame, self-reproach and self-depreciation with lifelong inefficiency. The child will rather be met halfway and in a franker spirit will discover the effective and useful path out into social cultural behavior where his forces receive actual opportunity.

There is definite work along each stage of the child's development. Each libido trend, partial impulse, is a thing of actual value, of tremendous power for good or evil. It already in the child's life makes for his constructive happiness and satisfaction or builds up a long train of morbid and asocial reactions, producing destructiveness in society and unsuspected misery on the part of the child. Parenthood looks to the physician to counsel and direct training as well as to meet the individual manifestations that are called disease, abnormalities, bad habits, anomalies. Just so the physician cannot meet these intelligently and effectively unless he looks to psychology. And it must be this deeper psychology which taps the dynamic forces that are working through the child's development. There is the immeasurable past to be considered in which lies the psychical history of the race, of which the child is only one transitory part. Out of it stream the forces which make even the small child what he is and condition his striving, his pleasure seeking and the diverse and primitive ways it takes, as well as the strength and value of the adult aims toward which he is unconsciously building.

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